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VOL. 36 AUGUST 1951 FASC. 2

FURTHER CASES OF ABERRANT PANCREAS[1]

by

Lennart Kjellman

In a previous article an account was given of three cases of aberrant pancreatic anlage, two of which were located in the stomach and one in the duodenal bulb. No pathological confirmation was possible, as operation was not considered necessary. The roentgenological findings, however, amply justified the diagnosis.

The abnormalities observed were briefly as follows: a deformity in the contrast caused by a well-defined rounded shallow indentation of the mucous membrane, which on repeated examination appeared to be quite benign, with a crater-like hollow in the centre, which filled with contrast medium. Leading from this cavity, were fine canals, also filled with contrast medium, of varying width and appeared to be capable of contraction; these terminated peripherally in small club-shaped processes and in fact had the general appearance of glandular ducts. In other words the condition was of a congenital nature.

After this article had gone to press a note was added to the effect that another case with similar roentgenological findings had been operated upon and the diagnosis confirmed pathologically. An account of this case is given below, together with three further cases of aberrant pancreas.

Case 1 (Figs. 1—3). A male, now 36 years old, who, in 1938, had begun to experience aching pain in the mid-epigastrium, particularly at night, but without vomiting or indisposition. He could tolerate all types of food. The symptoms diseappeared later and returned in 1943. A roentgen examination carried out at another hospital in December of that year and also at a later date showed: some irregularity of the mucous membrane of the stomach, with signs of increased secretion; marked ulcerative deformity of the duodenal bulb with deep involutions on the major and minor curvatures; further, situated distally on the anterior wall close to the minor curvature an ulcer-like niche about the size of a bean. After four weeks treatment a niche about the size of a peppercorn remained,

[1] Submitted for publication, April 27, 1951.

From the Roentgen Department (Director: Professor ÅKE ÅKERLUND), Karolinska sjukhuset, Stockholm, Sweden.

Case 1.

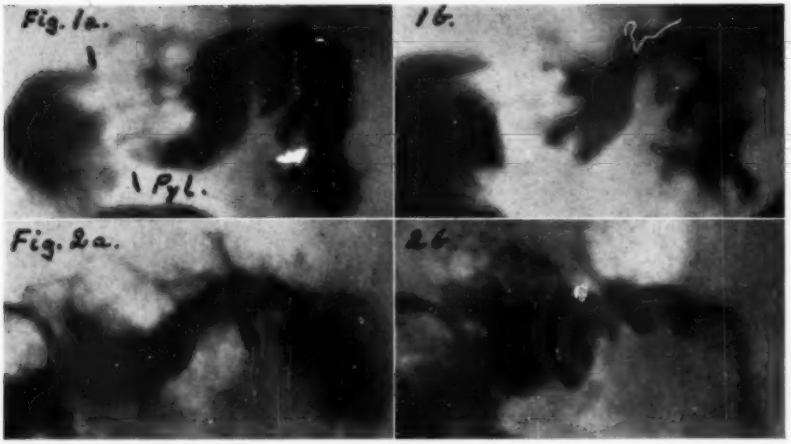

Fig. 1 a—b. Serial roentgenograms, 1944, showing a deformity of the bulb and irregularity of the ducts.
Fig. 2 a—b. Reexamination, 1950. The same deformity but only an irregular duct opening.

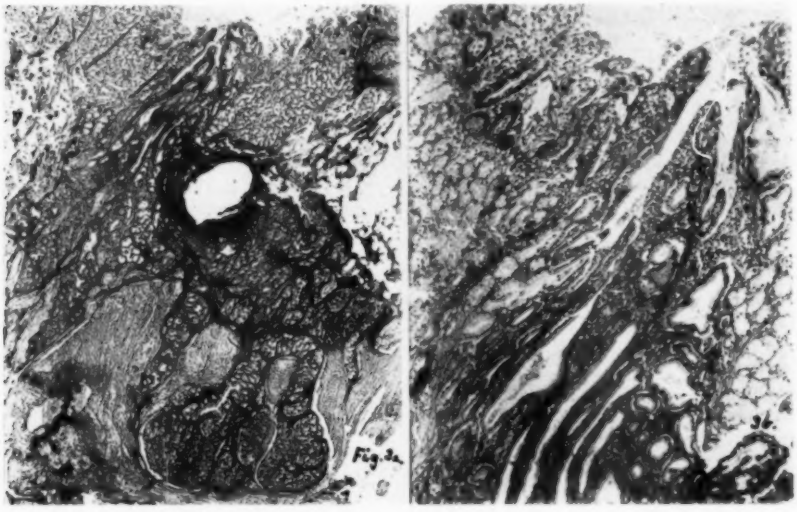

Fig. 3 a—b. Photomicrographs. a, 7 × and b, 50 ×.
Staining: van Gieson.

and this after a further two weeks appeared to have regressed. In spite of diet, there was a further exacerbation in 1944, when a niche the size of a peppercorn was again observed. Two months later, it seemed to have diminished in size. No more pain was experienced in the next two years, but thereafter, especially in the autumn and spring, symptoms recurred similar to those already described. In July 1950 the epigastric pain was more pronounced than it had been previously. A roentgenogram in August showed a deformity of the duodenal bulb with a probable active ulcer in the distal part.

The patient was therefore referred to the Surgical Clinic of the Karolinska sjukhuset for operation. In September a gastro-duodenal resection was performed because of the above mentioned diagnosis of a chronic duodenal ulcer. The technical difficulties of the operation proved to be surprisingly great, partly because the patient was rather fat, and partly because the duodenal bulb was fixed to the pancreas by adhesions and could not be drawn out, but was, with some difficulty, removed piecemeal. The pancreas, however, appeared to be uninjured.

Microscopic examination was made of three pieces of tissue about the size of almonds, which were removed from the duodenal bulb. They were partly covered with mucous membrane, but it was impossible to reconstruct their topography. Several sections showed a duodenal wall with islands of pancreatic tissue consisting of excretory parenchyma and ducts running between the smooth muscle bundles. There was no ulcer and no evidence of malignant tissue. The pathological diagnosis was: Aberrant pancreatic tissue.

On inspection of the roentgen films received from the other hospital confirmation of the deformities in the distal part of the duodenal bulb described in the reports was made. Furthermore, in several of the roentgenograms taken on different occasions, two ducts could be observed, which were at least $^1/_2$ cm long and filled with contrast material. They proceeded from the mucous membrane of the posterior wall of the duodenum and there was some indication of branching at their peripheral terminations. The motility of the mucous membrane in this region, appeared, however, to be unaffected, and in serial films the ducts appeared to vary in width and contain a varying amount of contrast filling; obviously owing to different stages of contraction. At other times only a short funnel-like bulge of varying character appeared, corresponding to one of the openings of the canals, and it was perhaps this finding that was taken for an ulcer niche, although such a lesion is scarcely apparent in the films. In fact, roentgenograms over a period of years showed a consistent picture of duct-like processes filled with contrast medium indicative of aberrant pancreas tissue.

Case 2 (Fig. 4). A woman 50 years old with no previous history of stomach complaint had been nervous for a month, while her mother was ill, and troubled with regurgitant sensations in the epigastrium and occasional vomiting. No heartburn or acid eructation. No abdominal pain. No abnormality of the faeces. Nothing pathological palpable in the abdomen.

Roentgenological examination of the stomach showed only a moderately accentuated outline of the mucosa. However, situated on the anterior wall of the duodenum, just at the base of the bulb, was a deformity half the size of a hazelnut, caused by an evenly rounded indentation of the mucous membrane with a central funnel-shaped excavation from which there ran a fine canal, 1 cm in length, terminating peripherally in a club-like swelling. In a series of roentgenograms the changes appeared to bear a relationship to the movements of the anterior wall, and the canal varied in shape and in the degree of contrast filling. The outline of the mucous membrane within the duodenal bulb was also rather accentuated. There was no delay in the emptying time. The findings were compatible with a diagnosis of aberrant pancreatic anlage.

Case 2.

Fig. 4 a—b. Serial films at the first examination, 1950, demonstrating the constant filling defect of the bulb and a fine central duct.

On further examination, three weeks later, the patient, having received sedative treatment, was quite free from symptoms. The roentgenogram still showed the contrast defect in the anterior wall of the duodenal bulb and the presence of the fine contrast-filled canal, unchanged in appearance. This gave further support to the assumption that this was a case of aberrant pancreatic tissue.

Case 3 (Figs. 5—6). A conscript 21 years old with no previous stomach complaint. During training he stumbled and fell, and a hand grenade pin which was stuck in his belt was driven into the pit of his stomach without, however, causing a wound. He did not lose consciousness, but immediately after the accident he suffered severe pains in the epigastrium and for a short time had difficulty in breathing. Repeated vomiting occurred during transport to hospital. On admission to the Karolinska sjukhuset the patient was in a shocked condition and there was a tender region in the epigastrium but no signs of peritonitis. There was repeated vomiting and haematemesis. A blood-transfusion was carried out and morphine administered. Roentgenological examination of the abdomen revealed nothing pathological. The condition improved progressively, and after five days only slight tenderness remained.

On the eighth day, when a roentgen examination of the stomach was made, the tenderness on palpation had disappeared. There was significant hypersecretion, and on the greater curvature there appeared an evenly rounded indentation of the mucous membrane into the lumen of the stomach with a central funnel-shaped cavity from which there ran a fine canal, about 1 cm in length. This was visible in two of the films. The abnormality was displaced by movements of the stomach wall, and thus caused no interruption of the peristaltic waves. All these facts combined to support the conclusion that

Case 3.

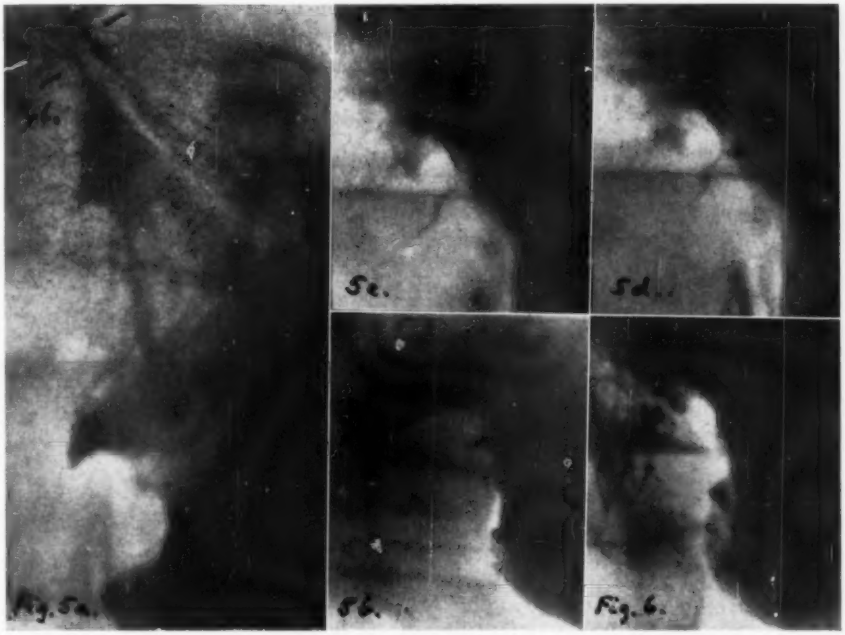

Fig. 5 a—d. Roentgenograms showing a spherical defect with a cavity and a fine duct.
Fig. 6. Control three weeks later giving the same picture except for the duct filling.

this was an aberrant pancreatic anlage. The patient was discharged the next day, symptom free.

A few days after arrival home the patient experienced general abdominal discomfort with heartburn, frequent indisposition and vomiting $^1/_4$ hour after meals. He was readmitted to hospital a week later, whereupon the symptoms promptly yielded to antispasmodics and did not recur. The roentgenogram was unchanged, except for the fact that there was no contrast material in the duct on this occasion.

Case 4 (Figs. 7—11). This case has kindly been placed at my disposal by Docent IVAR PALMER, Director of Surgical Department I, Södersjukhuset, Stockholm. It concerns a 26-year-old man, who in 1943, at the age of 19, had been operated upon for perforated ulcer of the stomach, gastrorrhaphy being performed. In 1944 typical symptoms of duodenal ulcer arose. According to the reports, the roentgenograms showed signs of gastritis and a severe, persistent deformity of the duodenal bulb, with deep involutions from the major and minor curvatures and a central niche about the size of a pea. After five weeks hospital treatment the lesion healed.

The following year the patient suffered from renewed discomfort of an ulcer-type. Roentgenological examination at the Karolinska sjukhuset showed an irregular outline

Case 4.

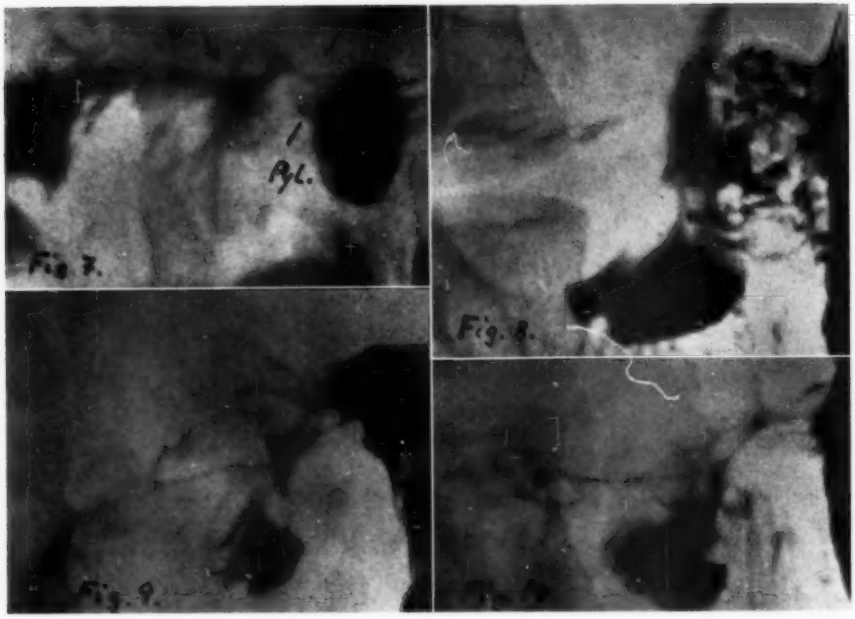

Fig. 7. Roentgenogram in 1944 demonstrating a deformed bulb with an ulcer (U).
Figs. 8—10. Reexaminations in 1945, 1949 and 1950 showing the same deformity of the bulb but no ulcer.

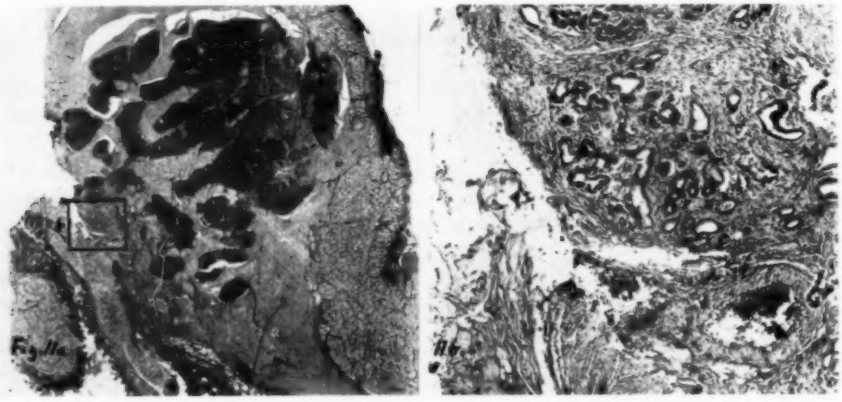

Fig. 11 a—b. Photomicrographs. a, 7 × and b, 45 ×.
Staining: Ehrlich — Htx — Eosin.

of the mucous membrane of the stomach and signs of hypersecretion. There was a pronounced deformity of the duodenal bulb, but no definite niche could be observed. On several occasions in the following year the patient experienced only slight epigastric discomfort which readily responded to antacid treatment.

In 1949 the patient complained of slight abdominal discomfort over a period of two weeks and then suddenly had tarry stools and felt weak. There was no pain at this time. On admission to hospital the presence of blood in the stools was verified. A blood examination showed no signs of anaemia. The patient underwent a four weeks course of ulcer treatment and during the whole time was symptom-free. After a week there was no longer any blood in the faeces. Roentgenological examination one week after admission showed no pathological abnormality in the stomach. There was an extreme deformity of the duodenal bulb, but no definite niche.

After discharge, the patient was treated in the Out-Patient Department. A year later symptoms of an ulcer-like character were still present and the roentgenograms showed the existence of a central niche in the severely deformed duodenal bulb. The patient was recommended for operation and admitted to the Södersjukhuset, where a gastric resection was performed.

Pathological report: No obvious macroscopic or microscopic evidence of an ulcer niche or cicatrization. In a preparation from the lower margin of the resected tissue, however, there was some cicatricial formation in the sub-mucosal connective tissue. Islands of considerable size which were clearly aberrant pancreatic tissue were also visible just beneath the mucous membrane and in the musculature. The mucous membrane of the stomach was moderately inflamed but there was no evidence of malignancy.

Thus the microscopic examination showed, as in the first case, that the aberrant pancreatic tissue consisted of excretory parenchyma as well as ducts, some of which, moreover, broke through the mucosa and opened into the lumen of the intestine. The demonstration of such excretory ducts also provides an explanation for the canals observed in the roentgenograms; particularly a retrograde filling is a plausible possibility in the absence of any sphincter.

In view of this latter case, it now seems very probable that the interpretation of the roentgenological findings as summarized from the earlier cases, was correct. The same circumstances also provide strong support for the diagnoses in cases 2 and 3, in which ducts ran from the centre of the indentations in the mucosal outline.

The generally accepted opinion is, however, that roentgenological findings cannot definitely establish the presence of aberrant pancreatic anlage, even though such a malformation occasionally may have been suspected, and that the diagnosis of a polyp is the nearest that the radiologist may come to a definite diagnosis. An abnormality located in the duodenal bulb may also often be mistaken for an ulcer, as occurred in the last case, where two weeks after roentgen examination, no niche was observed microscopically, but where there were visible islands of aberrant pancreatic tissue. In contrast to the patent ducts of case 1, this case revealed no channels opening on to the surface of the mucosa, and thus it was theoretically impossible to fill them with contrast medium.

However, it is clear that the roentgenological findings of filling defects which from all evidence appear to be benign and which, moreover, contain contrast-filled canals of variable width, give such an unequivocal picture that they permit a diagnosis of aberrant pancreatic tissue.

SUMMARY

An account is given of four cases of aberrant pancreas, two of which were confirmed. The malformation was in one case located in the stomach, and in the other in the duodenal bulb. The occurrence in roentgenograms of ducts filled with contrast medium, is pointed out as having considerable diagnostic significance.

ZUSAMMENFASSUNG

Es erfolgt ein Bericht über 4 Fälle mit aberrierendem Pankreas, von denen 2 bestätigt wurden. In dem einen Falle fand sich die Missbildung im Magen, in dem andern im Bulbus duodeni. Es wird darauf hingewiesen, dass kontrastgefüllte Gänge, die im Röntgenbild zur Darstellung kommen, dafür diagnostisch kennzeichnend sind.

RÉSUMÉ

Relation de quatre cas de pancréas abérrant dont deux ont été confirmés. L'anomalie se présentait une fois au niveau de l'estomac et dans les autres au niveau du bulbe duodénal. L'apparition de conduits remplis de contraste sur la radiographie est considérée comme d'une grande importance pour le diagnostic.

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FROM THE ROENTGEN DEPARTMENT (DIRECTOR: PROFESSOR T. DALE) OF THE UNIVERSITY HOSPITAL, OSLO, NORWAY

FIBROUS DYSPLASIA — FIBRO-OSTEOMA — OSTEOMA OF THE FACIAL BONES AND THE SKULL[1]

by

Andreas Hobæk

When, in 1937, ALBRIGHT and his associates (1) published the first cases of the disease now usually called Albright's Syndrome, the authors termed the bone manifestations of this disease »osteitis fibrosa disseminata». Thus the problem of having to deal with three different forms of »osteitis fibrosa» in clinical and roentgenological nomenclature arose: v. Recklinghausen's »osteitis fibrosa cystica generalisata» (hyperparathyroidism), the new disseminate form, and the so-called »osteitis fibrosa localisata».

The term »osteitis fibrosa» has by priority been attached to v. Recklinghausen's disease, hyperparathyroidism, in clinical terminology. Being a general disease it will necessarily affect the whole bone system: it is therefore illogical to maintain a disseminate, and still more a local type, of »osteitis fibrosa» in clinical terminology.

The coining of the term polyostotic fibrous dysplasia of bone by LICHTENSTEIN (2) as a substitute for »osteitis fibrosa disseminata» the year after Albright's report was published has contributed somewhat to a clearer conception of the problem. »Osteitis fibrosa» is, however, originally and principally a pathologico-anatomical term. By far the greater number of pathological conditions affecting the bone system will, owing to the structural peculiarities of this tissue, lead to a localized osteolysis, regardless of the nature of the pathological condition from which it has originated. As far as is known the breaking down process will always take place by osteoclasis. The tissue which fills up the empty spaces after the bone has been destroyed, may be of such a histological specific type that it is relatively easy to identify, as in primary or

[1] Submitted for publication, Jan. 16, 1951.

metastatic malign tumors or in acute inflammatory conditions. Although this replacement tissue in a number of diseases shows mainly a uniform histological structure, it must be assumed that they have a totally different etiology. The histological appearances are characterized by connective tissue, which is more or less rich in collagen fibrils containing a varying number of fibroblast- to osteoblast-like cells, often with areas of osteoid tissue, and a greater or smaller amount of giant cells of osteoclast type (»cellules à myeloplaxe»). Numerous trabeculae of newly-formed or older bone with borders of osteoid tissue or surrounded by osteoblasts in pallisade arrangement are, as a rule, seen in the connective tissue.

The immature bone seems rarely or never to develop into any functionally fully normal, architectonically arranged structure. It remains primitive. The development of this »osteitis fibrosa» tissue must be assumed to be the unspecific, reparative response of the organism to damage, which may be due to a number of different causes. In this tissue we find the main features of normal histogenesis of fibrous bone, but development is generally halted before the stage of functionally normal, mature, lamellar bone is reached.

To the group of diseases with this mainly common histological character, belong the bone changes in primary and secondary hyperparathyroidism, osteitis deformans Paget and polyostotic fibrous dysplasia, all of them diseases with a generalized or disseminate distribution in the bone system. The diagnosis may, as a rule, be made without any particular difficulty on the characteristic roentgenological appearances and significant clinic-biochemic data. But, with the exception of the older, well-developed forms of osteitis deformans Paget where the intense variation in the degree of destruction and new formation of bone within neighbouring areas of the same bone section, gives the histological pattern a typical »mosaic» structure, it is very difficult to differentiate the diseases in the »osteitis fibrosa»-group from each other on the histological picture alone. There are variations, but no constant and significant differences in the pictures. Such variations are of a quantitative rather than of a qualitative nature.

As regards diseases which are grouped under the comprehensive diagnosis of »osteitis fibrosa localisata», the absence of characteristic polyostotic or generalized spreading in the skeleton and the significant biochemic data, will make an exact diagnosis still more difficult. This group comprises supposed monostotic forms of fibrous dysplasia, osteitis deformans Paget and fibro-osteomas. The central fibromas of the jaws and possibly other related diseases also probably belong to this group.

It is assumed that the histogenesis of fibrous bone takes place according to the following scheme:

1st phase:
fibroblasts
+ collagene fibrils
+ interfibrillar substance
↓
»osteoid»
↓
»ossein»
↓ ↙ apatite (hydroxy- or carbonate- or both)
Immature, coarse-trabecular, not lamellated bone.

2nd phase:
This primitive, immature bone is gradually removed by osteoclasis, — and the developmental steps repeated, this time with mature, lamellated, functionally organized bone as the result.

Histologic sections from »osteitis fibrosa»-tissue may show all the developmental stages of the first phase of this scheme. But after the destruction of normal bone by an »osteitis fibrosa» disease, a local factor seems to be lacking, a factor which in the physiological development of bone catalyses the final steps in the maturation of this tissue, and ends with the building up of trajectors. All the developmental stages of maturation may be encountered, but never fully mature, functionally arranged bone. Even in osteitis deformans, where the bone in affected areas comes nearer to maturity than in the other mentioned diseases, the bone structure never becomes normal. The bone changes in hyperparathyroidism which disappear after the extirpation of adenomas in gland. parathyreoidea, are possibly an exception to this rule, as evidently normal, mature bone is developed during the reparation phase.

The facts mentioned above indicate that »osteitis fibrosa»-tissue must be considered an unspecific reparative tissue, originating in bone marrow from mesenchymal structures which possess the ability to complete the first phase of the normal histogenesis of bone. But the lack of one or more unknown factors wholly or partly prevents the completion of the second phase of normal histogenesis ending in mature bone. This or these factors must evidently be local, as the physiological osteolysis and osteosynthesis of bone goes on undisturbed in the non-affected bone areas.

On the basis of what has been said above it will be clear that the histological-morphological picture does not permit any exact differential diagnosis between diseases within the »osteitis fibrosa»-group, and it is probable that only a detailed study of the histochemistry of the cell-structures will present opportunities for a clear differentiation.

»Osteitis fibrosa localisata» of the facial bones and the skull now seems in American literature to be regarded as identical with fibrous dysplasia (JAFFE (3), PUGH (4), SOSMAN (5)), or a variant of this disease (SCHLUMBERGER (6)). In European literature LIECHTI (7) has held the same view.

Table I

Situation	Number of cases	Sex ♀	Sex ♂	Age	Number of cases operated	Histological diagnosis
Maxillae and zygomatic bones	12	7	5	11—43	5	Fibro-osteoma .... 3 Osteoma ..... 1 Giant cell t. 1
Mandibulae	4	1	3	21—27	3	Fibro-osteoma 3
Ethmoid bones	5	2	3	15—57	3	Osteoma ..... 2 Osteo-fibroma 1
Frontal Sinuses	21	8	13	18—39	4	Osteoma 4
Basis Cranii	3	2	1	15—62	0	
Cranial Vault	7	4	3	20—45	1	Osteoma 1

EDEN (8) and BILLING & RINGERTZ (9) maintain that fibro-osteomas, including the more diffuse forms, and osteomas, only represent different stages of maturation of the same disease, and prefer to comprise them under the term fibro-osteomas. These authors do not discuss the cranial localization of fibrous dysplasia, but hold that the greater number of cases of leontiasis ossea, and therefore probably fibrous dysplasia, may be designated fibro-osteomas of a diffuse type. TRAUNER (10) who, like BILLING & RINGERTZ, doubts the tumor character of this disease, chooses the more neutral term »hyperostosis». AXHAUSEN (11) seems to regard »osteitis fibrosa» in the jaws as an expression of osteitis deformans Paget (»Kiefer-Paget»).

I shall now discuss the roentgenological diagnosis of different diseases of an »osteitis fibrosa» character situated in the bones of the face and skull, in connection with the report of 52 cases that have been examined at the Roentgen Department of The University Hospital in Oslo from the year 1932 to 1949. A number of osteomas have been included in the series for reasons which will be apparent from the discussion which follows. Table I gives a survey of the situation of the affection, age and sex of the patients, number of patients operated upon and the histological diagnosis. The age stated refers to the time of the first roentgen examination. The first clinical symptoms often dated back many years, even decades, before that time.

It will be seen from table I that surgery has been performed and histological examination carried out in only 16 cases. Most of the osteomas in the frontal sinuses have been discovered by accident and have not presented indications for operation. No reason for surgical treatment was apparent in the other cases, by reason of the fact that the situation of the lesion would have rendered a radical operation unnecessary muti-

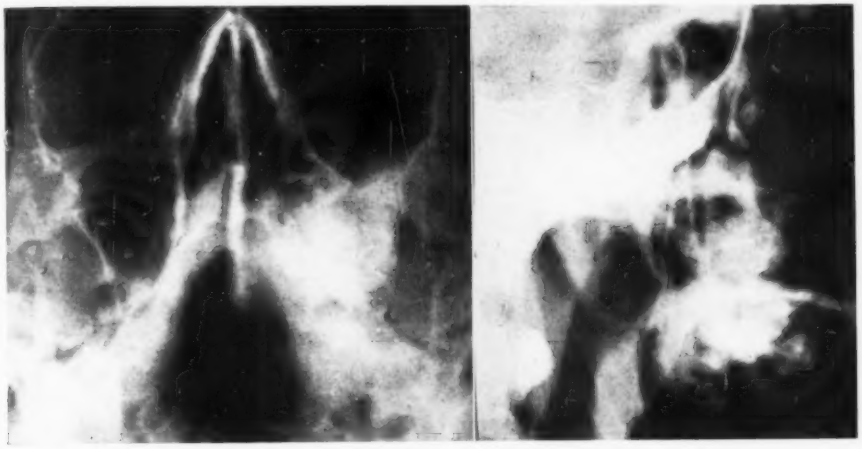


Fig. 1 a. Male, 43 years.

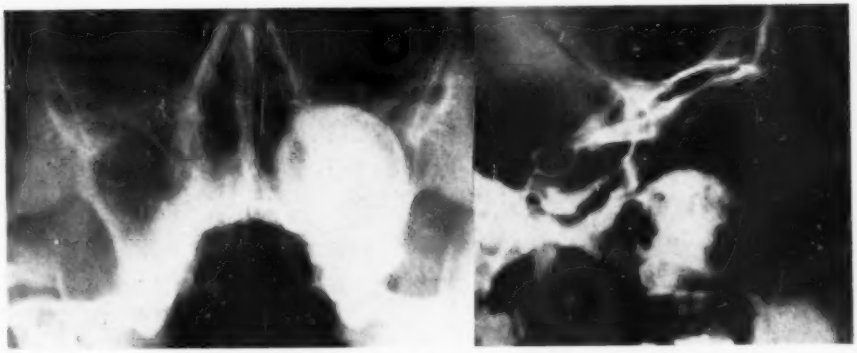


Fig. 1 b. Female, 29 years.

Fig. 1. Fibro-osteomas of maxillae.

lating or risky. In some of the cases the process seemed to be stationary and cosmetic considerations did not justify operation.

Fibro-osteomas (ossifying fibroma — osteo-fibroma — hyperostosis) are most frequently situated in the jaws, especially the upper jaw and antrum Highmori. They often seem to originate in the tuber maxillae or processus alveolaris. SHERMAN & STENBERGH (12) and PHEMISTER & GRIMSON (13) emphasize that »ossifying fibroma» ought to be segregated from the other forms of »osteitis fibrosa» in the same situation. According to SHERMAN & STENBERGH »ossifying fibroma» is monolocular, compa-

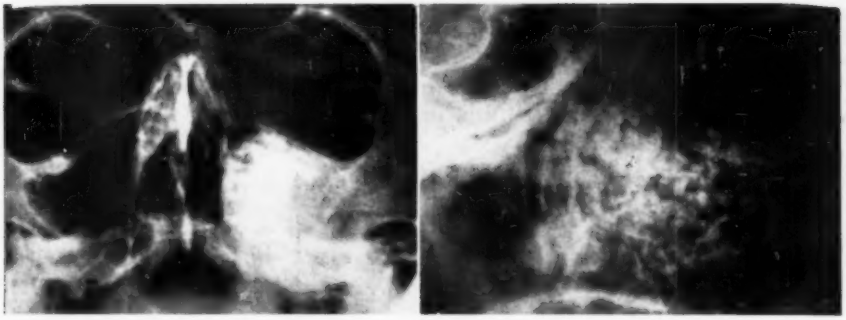
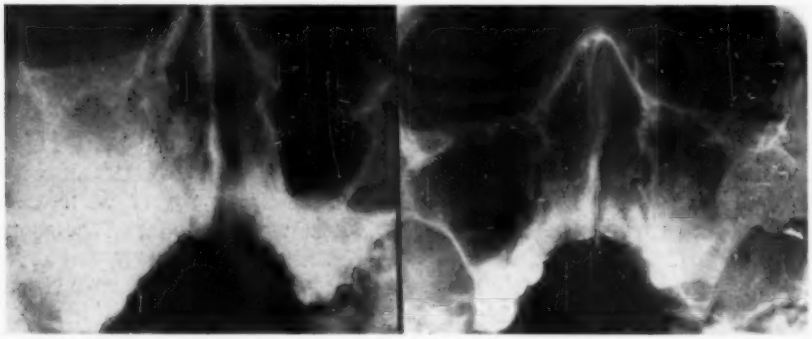


Fig. 2 a. Fig. 2 b.

Fig. 2. Fibro-osteoma. Female 52 years.



Male 17 years. Female 18 years.

Fig. 3. Fibrous Dysplasia of maxilla and zygomatic bone.

ratively well-defined, dates back to childhood or puberty and shows no histologic specificity.

The pathological-anatomical diagnosis in three of the five operated cases was fibro-osteoma, in one case osteoma and in one case giant cell tumor. Both BILLING & RINGERTZ and SOSMAN mention »degenerative» changes in fibro-osteomas with the appearance of localized areas of giant-cells. The diagnosis of »giant-cell tumor» in such cases is not unusual.

In eight of the cases the affection in the maxilla had a relatively well-defined border with a partly corticalis formation and a round to oval shape as seen on roentgen examination. Such cases (fig. 1) may show a quite pronounced sclerosis, rarely or never homogeneous, but with greater or smaller irregular less dense areas. In two of the cases the sclerotic areas were small and tightly packed so that the affection gave an impression

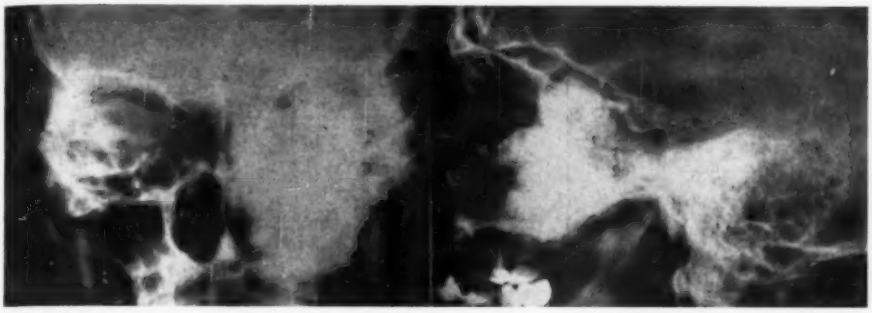


Fig. 4 a. Fig. 4 b.

Fig. 4. Osteoma. Female 29 years.

Fig. 5. Fibro-osteoma mandibulae. Male 27 years.

Fig. 6. »Pseudo-Paget». Male 25 years.

of granulation on the roentgenographs (fig. 2). RUPPE (14) has described »osteitis fibrosa» of this type in the jaws. In France the maxillar situation of »osteitis fibrosa» is usually called »maladie de Charles Ruppe».

Three cases showed less well-defined borders and spreading of the affection to the os zygomaticum and orbita (fig. 3). There was not such a profound sclerosis as in the previous cases, and the sclerosis gave the impression of being homogeneous. No biopsies were performed in any of these three cases. Roentgenologically, however, they are so different from the more well-defined fibro-osteomas that there may be some justification in classifying them as cases of fibrous dysplasia. They also seem to arise earlier in life. The growth seems to be very slow, but the process does not stop at the synostoses.

In one case the diagnosis was osteoma (fig. 4). The histologic sections

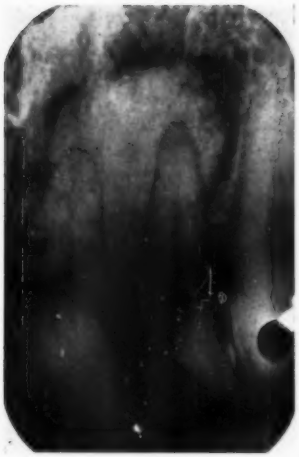


Fig. 7. Osteitis condensans.

showed closely packed, partly lamellar bone. There was a strikingly dense and homogeneous sclerosis compared with the other cases. The growth was lobulated and surrounded by a narrow translucent zone appearing as a border stratum to the surrounding normal bone. Thus the affection takes a more expansive character and the lobular shape suggests several expansion centres. Roentgenologically this tumor is so different from the previous two types that it seems reasonable to segregate it as a type of its own, an osteoma.

Three of the four mandibular affections have been operated upon. In these three cases the histologic diagnosis was fibro-osteoma. Radiologically the margins of the process in these cases were decidedly less well-defined than in the maxillar situation (fig. 5). In these three more diffuse affections the history suggested that the process started several years previously to the roentgen examination with pains and swelling in the lower jaw after tooth extraction. There was, however, at the time of the examination of the patients no clinical or histological signs of any inflammation. At this point, however, there is reason to refer to FUREDI's paper (15). She discusses the so-called »osteofibromas» in the maxilla and regards these as localized osteites of a sclerosing type; in 4 of her 14 cases she succeeded in demonstrating small bacterial foci in the histologic sections.

AXHAUSEN and SCHULZ (16) have described so-called »Pseudo-Paget» as an inflammatory condition of the lower jaw, with slow development and with periods of increase in the size of the swellings and redness of the skin over the affected jaw area. A hard thickening of the mandible of often quite considerable extent gradually forms. Histologically inflammatory changes can always be demonstrated. Fig. 6 shows a roentgenograph of the mandibula in a man, 25 years old, with this disease. During a period of about seven years a thickening of the right mandible had gradually spread towards the left. The swelling had increased slowly in size. At intervals of from one to two years marked soreness and increased swelling had occurred over periods of three to five weeks duration. There was never redness of the skin. No biopsy was performed. Roentgenologically, however, it is not possible to distinguish the picture presented by this process from a fibro-osteoma.

It may also be worth noticing that in four of the eight cases of maxillar affections of fibro-osteoma type reported above, there was a history of

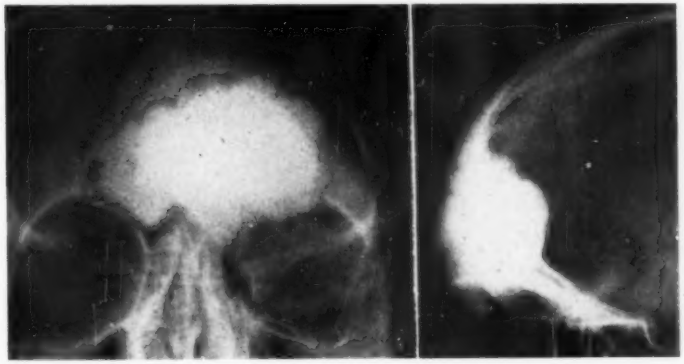


Fig. 8 a. Fig. 8 b.

Fig. 8. Large expanding osteomata of frontal sinuses. Male 18 years.

tooth extractions and subsequent inflammation of the maxilla on the side where the changes developed. It has never been possible, however, to demonstrate indisputable inflammatory changes in the histological sections that have been examined.

Localized osteitis condensans in the upper or lower jaws (fig. 7) is frequently found and is considered to be the result of chronic inflammation. It is most often found in edentulous jaws and is occasionally the cause of neuralgia. It is far from possible to succeed always in demonstrating inflammatory changes in the histological sections of such affections. The roentgenologic picture is characterized by more or less well defined osteosclerotic areas in the region around the apices of teeth. In cases of osteitis condensans, where it has not been possible to demonstrate histologically any sign of inflammation, one cannot immediately reject the possibility that the changes may have had an inflammatory origin. It must be assumed that these chronic osteites of productive type are caused by slightly virulent micro-organisms. It is not unreasonable to assume that some fibro-osteoma-like affections of the jaws have an inflammatory origin, even if the histological sections do not show any convincing signs of active inflammation.

The small osteomas in the frontal sinuses are relatively frequent. v. BAJKAY (17) has, in 1940, found 403 cases reported in the literature. Most of the frontal sinus osteomata in this series are accidental findings. They are in general of very little clinical interest. Only four out of twenty-one cases have been subjected to operation. In two cases small osteomas consisting of compact, lamellar bone were removed owing to persistent pain in the forehead. There were no signs of complicating sinusitis. Both patients have become free of symptoms after operation. The two other

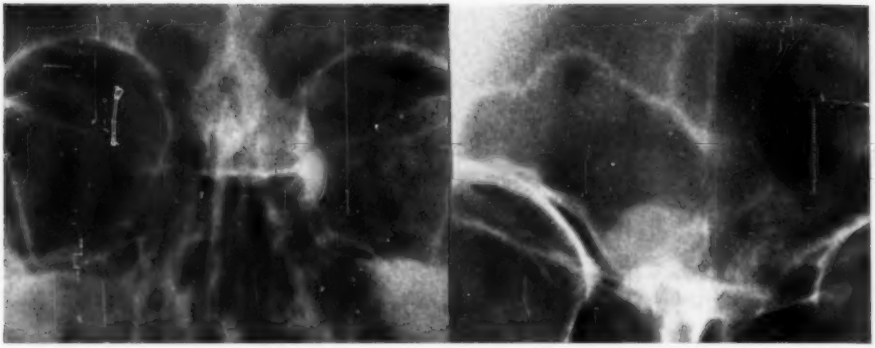


Fig. 9 a. Female 16 years.

Fig. 9 b. Male 24 years.

Fig. 9. Osteomas of ethmoid bone.

Fig. 9 c. Male 15 years (same Case as in fig. 10.)

cases operated upon are of greater interest (fig. 8). Here we had to deal with comparatively quickly growing tumors in young individuals. The tumors were large and had a distinctly lobular shape, dense sclerosis and a translucent border zone separating them from the normal bone. As suggested by CUSHING (18) they evidently most often originate in the os ethmoidale. In both cases reported here the tumor seemed to arise in the lamina cribrosa, but have been regarded as belonging to the frontal sinus osteomata by reason of the fact that the greater mass of the tumor is localized to these cavities. Compact mature bone with rudimentary Haversian systems were found histologically in these cases. The two tumors caused relatively marked pressure symptoms from the central nervous system.

The ethmoidal affections often lead to protrusio bulbi, and this usually is an indication for operation, even if the ocular power be not

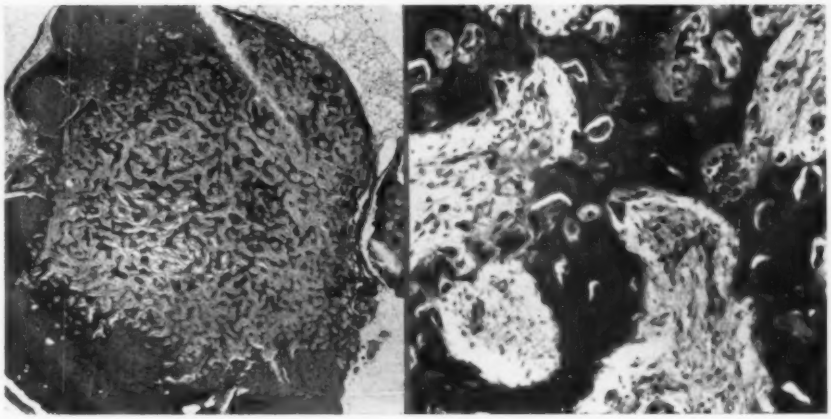


Fig. 10 a. Fig. 10 b.

Fig. 10. Osteoma of ethmoid bone. Male 15 years.

reduced. The histological diagnosis in two of the three cases operated upon was osteoma, and in the third »osteitis fibrosa» (fig. 10). Roentgenologically these tumors showed well-defined borders and a varying degree of sclerosis (fig. 9). They are apparently akin to the frontal sinus osteomata. It was not possible to demonstrate any cartilage in the sections.

Diffuse hyperostotic less well-defined affections of the basis cranii infiltrating several bones have been observed in three cases. Like the previously mentioned diffuse affections in the maxilla, these cases may very well be regarded as expressions of fibrous dysplasia. No other situations of fibrous dysplasia have been observed in these cases. The affections have been unilateral and mainly situated in the os frontale, and infiltrating the anterior cranial fossa and the os sphenoidale (fig. 11). Two of the patients were under twenty years of age, the third being 62 years old. All of them had been afflicted since childhood. None had been subjected to operation. The development of the disease evidently proceeds very slowly, often for decades. The thickening of the os frontale and other bones of the orbit not unrarely causes dislocation of the eye. This type of affection has not been observed in the cranial vault in this series. DENSTAD (19) has reported 3 cases of unilateral hyperostosis in the os frontale and the os sphenoidale of the same type as those included in the present series. These slowly growing diffuse hyperostoses were previously diagnosed as leontiasis ossea. As emphasized by PUGH it is probable that most of the cases of leontiasis ossea must be regarded as

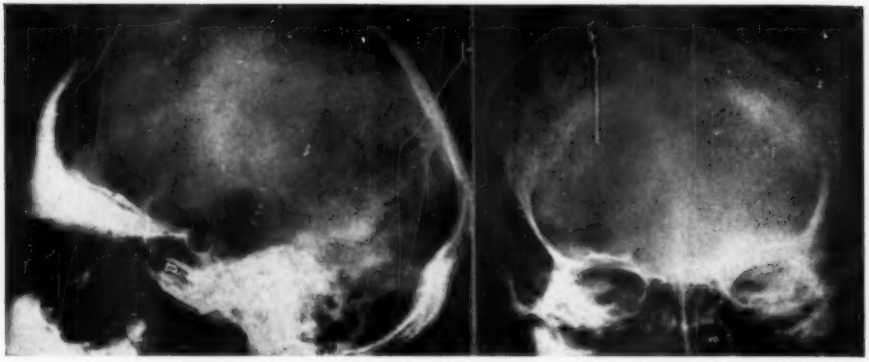


Fig. 11 a. Fig. 11 b.

Fig. 11. Fibrous Dysplasia. Female 15 years.

expressions of fibrous dysplasia, though a smaller number must be attributed to thickenings of the cranial bones caused by osteitis deformans. The possibility cannot be excluded that the etiology of hyperostoses of the cranial bones may in some cases be a mild inflammatory process, a kind of »creeping periostitis».

The seven cases of affection of the cranial vault in this series have the character of osteomas: pronounced sclerosis, translucent border zone and a slightly lobular shape. In the one case operated upon (fig. 12) bone of mature type was found on section. The growth is generally slow and expanding.

Osteomata are the most peculiar type of all bone tumors. EWING (20) emphasizes that it has never been possible to give a clear definition of the tumor type. The result of the growth of the osteoma is mature lamellar bone with more or less complete Haversian systems, but densely packed and without distinctly functional arrangement. It has never been demonstrated that the growth center of an osteoma lies peripherally, and that growth proceeds in such a manner that dense bone is built up in a steadily growing, centrally situated nucleus, while the growth expands peripherally. Growth seems to proceed periodically from one or more centres, and the potent bone-producing tissue appears to be able to remain at rest within the compact bone nucleus for a shorter or longer time, until a new growth impulse sets in. Already formed, compact bone (tumor-tissue) must then be broken down. After the growth impulse has ebbed out the expansion temporarily stops. VIRCHOW's observation is of great interest in this connection. He noted that cranial osteomas of spongy type became eburneous in the course of time, but could later

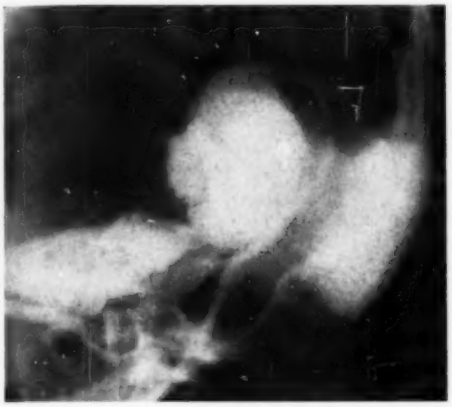


Fig. 12. Osteoma of temporal bone. Female 45 years.

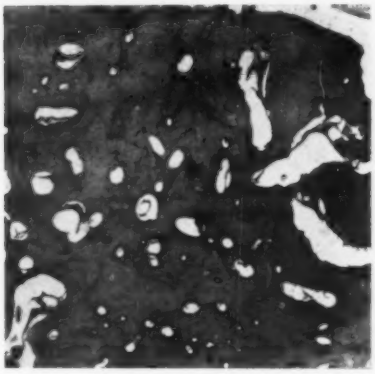


Fig. 13. Fibro-osteoma of mandibula. Compact bone with mosaic pattern.

again assume a spongy character. The lobular shape of the osteomas indicate that several growth centres are functioning at the same or possibly at different times. The histological picture of an osteoma during its expanding phase may suggest »osteitis fibrosa» in certain areas (fig. 10).

As previously emphasized, the ailments comprised in the group »osteitis fibrosa localisata» must be considered to be of different etiology. It must be assumed that the impact of each separate disease destroys the normal premorbid bone in a specific manner. This is not, however, clearly expressed in the morphological structure of the unspecific reparative tissue, which shows varying degree of maturation. It can overshoot the mark as in eburnised osteomata, or not go far beyond the stage of fibrous connective tissue as in central fibromata of the jaws. The breaking-down mechanism can evidently work parallelly with a constructive reparative process, as in »mosaic» bone. The actual process is registered both histologically and roentgenologically by the degree of the stage of maturation that is reached by the reparative tissue in each separate case. It is the very unspecific reparative tissue itself that binds the group together, and the histological and roentgenological appearances of the different types only reflect the difference in maturity of this tissue.

In fibro-osteomas with comparatively pronounced sclerosis more mature bone, laid down in a pattern suggesting mosaic, can sometimes be seen (fig. 13). TRAUNER maintains that this pattern is not identical with the »geordnete Mosaikstruktur» (SCHMORL) found in osteitis deformans Paget. But the way in which the bone is formed must, at any rate, be approximately the same. Peculiar to fibro-osteomas is apparently the

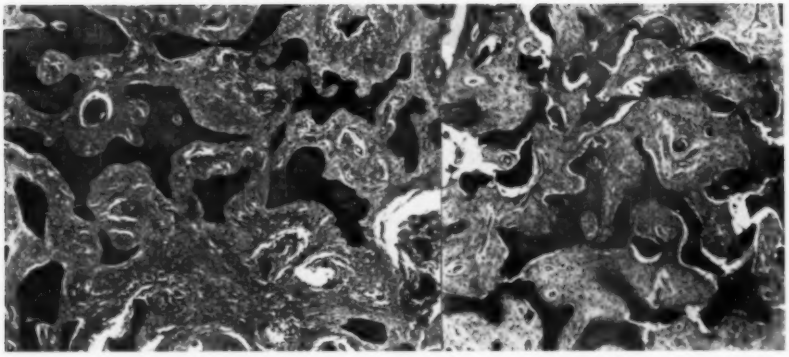


Fig. 14 a. Fibro-osteoma maxillae. Female 36 years.

Fig. 14 b. Fibro-osteoma mandib. Female 27 years.

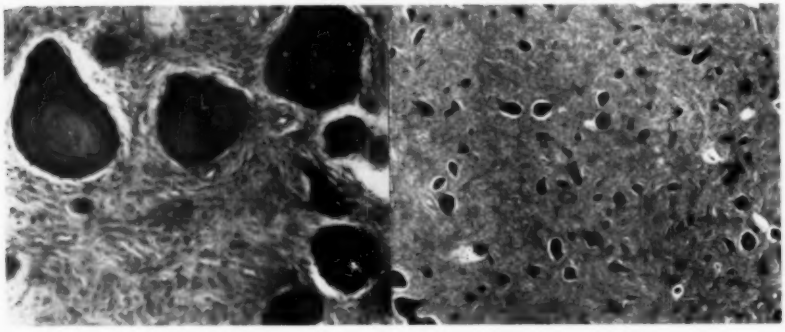


Fig. 14 c. Fig. 14 d.
Fibro-osteoma maxillae. Male 46 years. Psammoma-like bodies.

formation of small, round to oval, deeply stained bodies which may sometimes dominate larger or smaller areas of the histologic sections. (fig. 14). These bodies have a certain similarity to psammoma bodies, which also have the same concentric arrangement. A similar arrangement may also be seen in dentinomas. They are regarded as a result of osteoclasis of immature bone or as an expression of extremely primitive metaplastic bone formation. The bodies, however, occur only in isolated instances in fibro-osteomas and no decisive diagnostic importance can be attributed to them.

If fibro-osteomas, fibrous dysplasia and osteomata of the facial bones and the skull are to be regarded as separate pathologic entities, the main differential diagnostic difficulties will arise in distinguishing

them. The essential differential diagnostic points have been outlined in table II.

Table II

Type of lesion	Borders	Sclerosis	Principal situation	Age
Fibro-osteoma	Most often relatively sharp	Irregular Less marked to average	Maxillae, antra Highmori and mandibulae	At puberty or later
Osteoma	Sharp Extra-tumoral translucent zone. Lobular shape	Usually marked	Cranial vault. Paranasal sinuses	At puberty or later
Fibrous dysplasia	Definitely unsharp	Homogeneous Less marked to marked	Basis cranii (frontal and sphenoid bones). Maxilla and Zygomatic bones	Usually before puberty

The typical picture of osteitis deformans Paget of the skull will seldom offer any diagnostic difficulties. The age of the patient will be of certain assistance. The affection may have developed slowly, but never dates back to childhood or puberty. Most frequently the disease involves other parts of the skeleton. The demonstration of increased values of phosphatases in the blood serum supports the diagnosis. If the changes be situated partly or mainly in the jaws, the demonstration of widespread hypercementosis around the apices of teeth will support the diagnosis of osteitis deformans.

»Meningéome en plaques» is most frequently situated in the alae ossis sphenoidalis and the orbital roof. Women are mostly attacked by the disease, which is rare before the age of forty. Protrusio bulbi is often seen. The differential diagnosis in cases of monostotic fibrous dysplastic lesions occupying the sphenoid bone is difficult and sometimes impossible without biopsy. Encephalography will, as a rule, show dislocation or deformation of the ventricular system in cases of »meningéome en plaques», and cerebral angiography will often give useful informations.

Hyperparathyreoidism may offer considerable differential diagnostic difficulties in cases where the lesions are mainly situated in the bones of the skull and not obviously generalized. In such cases the diagnosis can only be made by a demonstration of the specific biochemic symptom-complex of hyperparathyroidism in the blood serum and urine.

Surgery, where indicated, is the only possible treatment of fibro-osteomas and osteomas. Recurrences are rare after radical removal. Billing & Ringertz and Ullik (23) have each described a case of fibro-osteoma with transition into sarcoma.

Indications for operation are seldom present in cases of fibrous dysplasia of the skull. Sherman & Stenbergh found that roentgenotherapy

effected no change. SOSMAN, on the other hand, maintains that he has observed retarded growth and increasing sclerosis after small doses. One of the patients in this series, a man 46 years old, who for more than ten years had had a slowly growing fibro-osteoma in the maxilla, received repeated roentgen treatment. Owing to the very slow growth, the effect was difficult to judge, but the neuralgiform pains of the patient temporarily disappeared.

Owing to the similarity of their histological structure there are many reasons for BILLING & RINGERTZ's proposal to regard the conditions described above as one pathological entity. In the series presented here, however, there seems to be a distinct enough roentgenological difference between the three types: the comparatively well-defined fibro-osteomata in the maxillae and mandibulae, the diffuse hyperostoses of the type fibrous dysplasia, and the osteomas of lobular shape with a translucent zone as a border stratum to the surrounding normal bone. It seems reasonable, at any rate for the time being, to discern between these three conditions, — but it must be admitted that they are not easily distinguishable.

SUMMARY

The author emphasizes that all types of »osteitis fibrosa localisata» of facial bones and the skull are characterized histologically by unspecific reparative tissue. This tissue, basically new fibrous bone formation, shows varying degrees of maturation, but no significant morphological differences. It is suggested on the basis of an analysis of the radiological examinations of 52 cases, that despite this histological uniformity, the discrimination between fibro-osteomas, fibrous dysplasia and osteomas of the facial bones and the skull should, for the time being, be maintained.

ZUSAMMENFASSUNG

Der Verfasser betont ausdrücklich, dass alle Arten von »Osteitis fibrosa localisata» der Gesichtsknochen und des Schädels histologisch durch unspezifisches Reparations-Gewebe gekennzeichnet sind. Dieses Gewebe, neu gebildeter fibröser Knochen, zeigt verschiedene Reife-Stadien, jedoch ohne besondere morphologische Differenzierungen. Auf Grund einer Überprüfung der radiologischen Untersuchung bei 52 Fällen, wird vorgeschlagen, dass trotz dieser histologischen Einförmigkeit die Unterscheidung zwischen Fibro-Osteom, fibröser Dysplasie und Osteom der Gesichts-Knochen und des Schädels zur Zeit noch beibehalten werden soll.

RÉSUMÉ

L'auteur insiste sur le fait que tous les types »d'ostéite fibreuse localisée» au niveau de la face ou du crâne sont caractérisés histologiquement par un tissu réparateur non spécifique. Ce nouveau tissu d'os fibreux se présente à des degrés de maturité divers, mais sans différences morphologiques appréciables. Il est cependant admis sur la base de l'analyse et de l'examen radiologique de 52 cas que, malgré cette uniformité histologique, la discrimination entre fibro-ostéomes, dysplasie fibreuse et ostéomes des os de la face et du crâne doit être maintenue.

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DIABETIC ARTHROPATHY[1]

by

Folke Knutsson

A grave condition of the joints occurring in diabetes mellitus and having the characteristics of a neuropathic arthropathy is described in the medical literature (BAILEY, ROOT, COZEN, MURI, FOSTER and BASSETT). The changes almost always are localized in the ankle-joints and the tarsi. The age incidence is between 30 and 60 years, and the patients have generally had diabetes for a number of years. The control and treatment of the disease has often been unsatisfactory.

The disease manifests itself as a more or less marked swelling around the ankle-joint and tarsus. The swelling often attains considerable proportions, but displays no pronounced inflammatory features: the skin shows no or negligible inflammatory redness whilst tenderness and local rise of temperature is slight or completely absent. The patients are able to support themselves on the foot and walk without pain. There is thus considerable discrepancy between the objective character of the joint disorder and the insignificant subjective symptoms. In this respect the condition resembles the form of arthropathy which is seen in tabes and syringomyelia. It is characteristic of the disorder that it is consistently localized in the ankle-joint and tarsus, whereas arthropathy in tabes and syringomyelia is most often localized in the knee and hip-joint and only less frequently in the ankle-joint.

The roentgen appearances in diabetic arthropathy are characterized by destruction of the epiphyses with erosion of the joint surfaces. The surrounding soft tissues contain loose fragments of bone in the form of small splinters or granules. Sometimes diffusely scattered bone sand may be observed around the joint as a result of disintegration of the osseous matter. Restorative periosteal regeneration of bone and ossifications in the soft tissues around the joint are more or less marked. The skeletal

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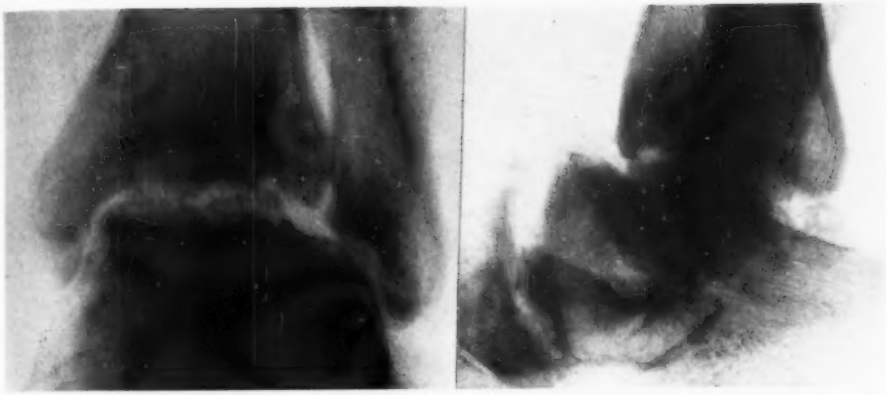

Fig. 1.

Fig. 1 (Case 1), 15.3.50: Extensive destruction with erosion of the joint surfaces. Alternating decalcification and sclerosis.

Fig. 2 (Case 1), 20.5.50: A group of small loose bony fragments (bone sand) is seen in the soft tissues in the anterior part of the ankle.

Fig. 2.

tissues retain their calcium content and decalcification occurs only in localized areas. The articular cartilages resist destruction at the commencement of the process but are progressly destroyed in the latter stages. In principle, therefore, the radiologic pattern is the same as that seen in tabes and syringomyelia.

The joint disease may thus, from the radiological standpoint, be assumed with some justification to be neuropathic. Both the appearance of the articular deformity and the absence of pain lend clinical support to this view. Syphilis, tabes and syringomyelia have been eliminated as causative factors in cases reported in the literature, and diabetes has been singled out as the probable etiology.

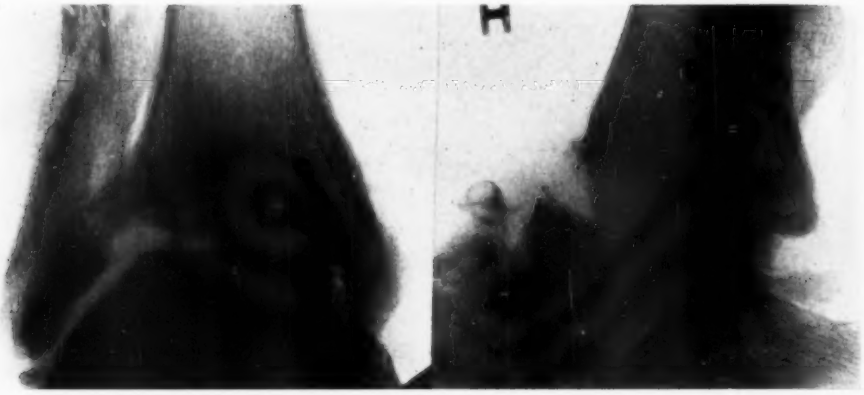

Fig. 3 (Case 1), 11.9.50: The joint surfaces are smoother and the sclerosis has increased. The roentgen appearances at this stage are characteristic of a trophic arthropathy.

Fig. 4 (Case 1), 24.10.50: The ankle is swollen and small pale scars following trophic sores are seen on some of the toes.

Pronounced diabetic neuritis with neuralgic pains is present in some cases; the patellar and Achilles' tendon reflexes are weak or are totally absent. This clinical picture is known by the name of diabetic pseudotabes. The patho-anatomical basis for the disease has been looked for in the neuritic degeneration that may occur in the peripheral nerves and in certain changes which also have been demonstrated in the dorsal fibres of the spinal cord.

Diabetic pseudotabes produces trophic changes in the skeleton and impairment of strength. A joint sprain may thus easily lead to skeletal and soft tissue lesions. The reduced or absent sensitivity to pain allows the patients to continue to use and injure the affected joint. This increases the destruction, gives rise to dislocations and counteracts attempts of the bone tissue to produce callus.

The trophic disturbance is probably the cause of the frequently occurring, very obstinate cutaneous ulcers of the feet which coexist with arthropathy in some cases.

Case 1. (Vfa 1458/50) Figs. 1, 2, 3 and 4.

Female; born 1921. Diabetes of long standing. She began to have trouble with the right ankle in 1945; this increased considerably in the Spring of 1950.

Fig. 5 (Case 2), 5.4.1951: A united fracture of the lateral malleolus and a pseudoarthrosis of the lateral malleolus are seen. The joint surfaces present circumscribed areas of destructions; the articular cartilages are intact.

In March 1950, on examination the ankle-joint was considerably swollen. It was but slightly tender and the patient could support herself and walk on the foot. There were also small trophic sores on some of the toes. The swelling continued to increase in size and in April was punctured and 15 ml of a sanguinolent fluid withdrawn. The punctate was devoid of tubercle bacilli. Aureomycin treatment was commenced and some improvement noted: the joint became absolutely painless but the swelling remained. After incision in June 1950 large quantities of greyish-yellow granular masses were removed. Joint capsule biopsy revealed: unspecific granulation tissue, no evidence of tuberculosis or a tumour. A guinea pig test of the joint fluid was negative for tuberculosis.

The Wassermann reaction of the blood was negative.

August 1950, examination of nervous system. Definite disturbances of sensation were present high up the leg, but these vanished two months later. The ankle joint was still swollen, but the trophic sores had disappeared.

Case 2. (A. S. U—a 16966/50) Fig. 5.

Female: born 1923. Grave diabetes of serious nature from childhood. Had sprained the right ankle several times. It was very swollen on examination in April 1951, but the patient could support her weight and walk on the foot without pain.

Clinical examination revealed that the patellar and Achilles' tendon reflexes were absent. Marked loss of sensation to pain and touch was present in both legs and feet. The sense of vibration was impaired.

Case 3. (Vfa 6913/50) Fig. 6.

Male, born 1916. Treated at Södersjukhuset, Stockholm, October—December 1950. For ten years had suffered from severe diabetes, and in recent years increasing disturbances of sensation had developed in both feet; he had noted difficulty in distinguishing heat and cold and complained of »pins and needles» and of numbness. *Arterio-*

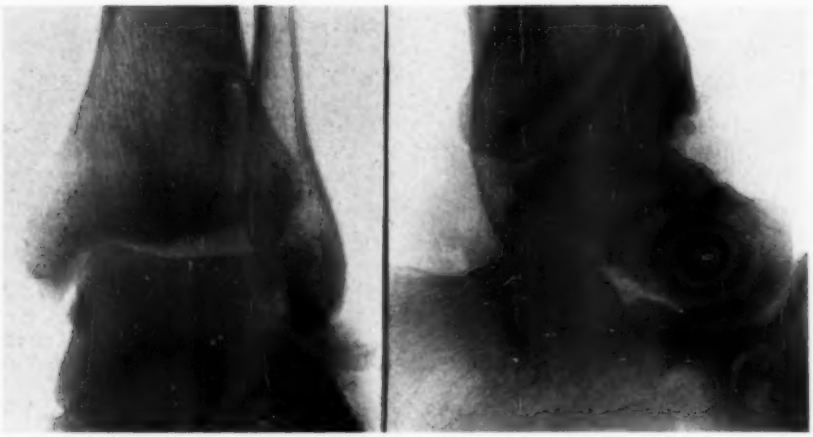

Fig. 6 (Case 3), 27.12.50: The radiograph reveals a slight subluxation of the talus of the ankle, a well-circumscribed defect in the lateral part of trochlea tali at the insertion of the capsular ligament, and marked posttraumatic ossifications in the adjacent soft tissues. The articular cartilages are normal.

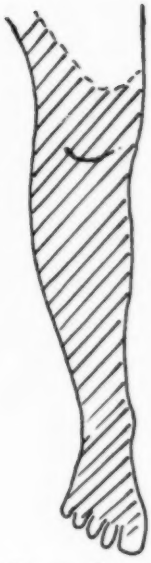

Fig. 7 (Case 4): The extent of absent sensibility to pain.

graphy of the left femoral artery revealed perfectly normal conditions in the thigh and leg. The Wassermann reaction of the blood was negative.

The left ankle-joint was twisted in 1949, but radiography disclosed no fracture. Since then, the swelling had not subsided. A clinical examination at the Norrbacka Institute, where the patient was treated in January 1951, showed that the ankle was very swollen but not painful or tender. The skin was slightly red and rather warm. The patellar and Achilles' tendon reflexes were absent and there was nil paresis. Pronounced loss of sensation to pain as well as to touch and temperature, mostly the latter, was present in the lower parts of both legs. The joint capsule of the ankle was examined by biopsy, and the histological examination revealed unspecific inflammatory changes.

Case 4. (A. S. U—a, K. Jansson, b. 19.3.88) Figs. 7, 8 and 9.

Male; born 1888. Treated for diabetes since 1932. In 1943 his legs began to go numb and ache. While in hospital in February 1944 it was found that both legs and feet exhibited impaired sensibility to pressure and touch, and that the sense of vibration was also reduced.

He was readmitted to hospital in February 1951. He then stated that he suffered from intermittent aches in the legs and had noticed some uncertainty in walking.

Four months before admission he sprained the left foot which became swollen. The swelling did not subside. The patient was able to walk on the foot all the time without feeling any pain.

Fig. 8 (Case 4), 20.2.51: The talus is subluxated forwards and a large defect embracing the anterior part of the tibial epiphysis suggests fractural sequelae. The adjacent soft tissues contain diffuse irregular ossifications. The joint surfaces present no signs of destructions and the cartilages are normal.

Fig. 9 (Case 4): Photograph showing the extent and degree of the swelling.

On examination he had a markedly swollen ankle which was slightly warm. There was neither tenderness nor pain.

There was absence of sensibility to pain in the legs or feet. Deep sensibility was absent and the sense of vibration impaired. Sensibility to touch was on the other hand intact. The Achilles reflex was absent, and the Wassermann reaction of the C. S. F. negative.

SUMMARY

Four cases of arthropathy in diabetes are described. All of them showed neuritic changes with impairment of sensibility to pain in the legs and feet. As in case reports in the literature the disorder was localized to the ankle-joints. Its radiological and clinical characteristics resemble those of a similar condition present in tabes; the cause is analogous, being in the nature of a neurogenic skeletal dystrophy accompanied by loss of sensibility to pain.

ZUSAMMENFASSUNG

Es werden 4 Fälle mit Artropathie bei Diabetes beschrieben, die alle neurotische Veränderungen mit herabgesetzter Sensibilität im Unterschenkel und Fuss aufwiesen. In Übereinstimmung mit den in der Literatur veröffentlichten Fällen war das Leiden im Fussgelenk lokalisiert. Die röntgenologischen und klinischen Veränderungen erinnern in ihrer Erscheinung an die Veränderungen bei der Tabes. Die übereinstimmende Ursache besteht in den neurogen bedingten trophischen Störungen im Skelettsystem und in der Abnahme der Schmerzempfindung.

RÉSUMÉ

Description de quatre cas d'arthropathie diabétique. Tous présentaient des altérations nevritiques sous forme d'amoindrissement de la sensibilité de la jambe et du pied. Comme dans les cas présentés par d'autres, le mal se localisait à l'articulation du pied. Son image radiologique et clinique rappelle fortement celle du tabès et les causes en sont analogues. Il s'agit de troubles trophiques du squelette de provenance neurogène accompagnés d'une diminution de la sensibilité à la douleur.

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AUS DEM RÖNTGENINSTITUT (LEITER: PRIVATDOZENT DR. E. A. ZIMMER)
DES KANTONSPITALS, FRIBOURG, SCHWEIZ

EIN FALL VON SYMMETRISCHER APLASIE ZWEIER LANGER RÖHRENKNOCHEN DER UNTEREN EXTREMITÄTEN, VERBUNDEN MIT VERSCHIEDENEN ANDEREN SKELETTMISSBILDUNGEN[1]

von

W. Güntert

Bei dem 10-jährigen Mädchen G. B. (Fig. 1), über das wir seiner seltenen Fehlbildungen wegen im Folgenden kurz berichten wollen, sind Kopf, Rumpf und obere Extremitäten normal geformt und dem Alter entsprechend entwickelt. Dagegen sind die Beine um mehr als halbe Beinlänge zu kurz, ungleich lang, und weisen keine beweglichen Kniegelenke auf. Die Weichteile deuten eine Gliederung in Ober- und Unterschenkel an, indem sie beidseits im proximalen Abschnitt der Extremität erheblich stärker ausgebildet sind als im distalen, wobei die Grenze durch eine vorne, medial und hinten deutlich erkennbare Furche markiert ist. Die »Oberschenkel» scheinen dabei auf etwa einen Fünftel ihrer normalen Länge reduziert, während die grazil gebauten »Unterschenkel» aspekt-

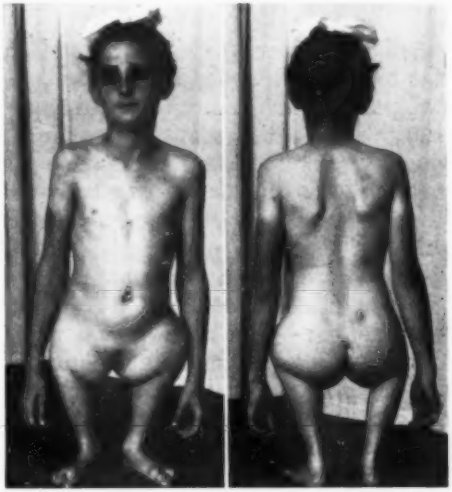

Fig. 1. Pat. G. B., 10-jährig, hochgradig verkürzte Beine ohne Kniegelenke.

[1] Vortrag an der Schweiz. Anatomentagung in Fribourg am 24.—25. Sept. 1949; bei der Red. am 28. März 1951 eingegangen.

Tabelle I

Bewegungsfähigkeit der Fuss- und Hüftgelenke.

a. Fussgelenke

Ausgangsstellung: Rechtwinkelstellung des Fusses.

	Pat. G. B.		
	Rechts	Links	Sollwert[1]
Dorsalflexion	0°	0°	25°
Plantarflexion	30°	45°	30°
Eversion	0°	15°	30°
Inversion	20°	10°	30°

b. Hüftgelenke

Ausgangsstellung: Rückenlage (beim Heben des Beines nach hinten Bauchlage), Beine möglichst aneinander, Mittellinie des Körpers fällt genau zwischen beide Beine. Hüftgelenke in 180°-Streckstellung.

	Pat. G. B.		
	Rechts	Links	Sollwert[1]
Beugung	90°	90°	100°—130°
Heben nach hinten	45°	45°	20°
Abduktion	35°	30°	70°
Adduktion	50°	55°	30°
Innenrotation	30°	35°	60°
Aussenrotation	110°	95°	40°

mässig ungefähr um einen Drittel zu kurz sind. Die Fussgelenke zeigen das typische Bild der VOLKMANN'schen Sprunggelenksdeformität, indem bei palpatorisch nicht feststellbaren äusseren Malleolen (Fibulaaplasie) die Füsse in Valgusstellung abgewichen sind. Die inneren Malleolen erscheinen kürzer und wohl auch etwas plumper als normal. Die Bewegungsfähigkeit der Fussgelenke ist eingeschränkt, wie aus Tab. I a hervorgeht. Das Auffälligste an den Füssen ist die Reduktion der Zehenzahl auf 3; ausser einer gut entwickelten Grosszehe sind beidseits nur zwei weitere Zehen vorhanden. Diese stehen in ziemlicher Entfernung von einander. Da zudem das Fussgewölbe erheblich eingesunken ist, möchte man — bei Vernachlässigung der sonstigen Fehlbildungen — von Pes transversoplano-valgus-Bildung sprechen (Fig. 2). Die Weichteile um beide Hüften, besonders um die linke, sind mächtig ausgebildet, was beim Stehen viel deutlicher in Erscheinung tritt als beim Liegen. Die Bewegungsmöglichkeiten der Hüftgelenke sind in Tab. I b zusammengestellt. Steht das Kind aufrecht, so reicht bei gestrecktem Arm die Spitze seines rechten Mittelfingers bis 8.5 cm, jene des linken bis 8.0 cm über den Fussboden. Die Distanz zwischen der Crista iliaca und der horizontalen Standfläche beträgt rechts 38 cm, links 36.5 cm. Es besteht

[1] DUBOIS-ZOLLINGER: Einführung in die Unfallmedizin, Bern, 1945.

Fig. 2. Pat. G. B. Pes transversoplano-valgus. Zehenzahl auf 3 reduziert. VOLKMANN'sche Sprunggelenksdeformität.

somit ein leichter Schrägstand des Beckens, der eine geringe skoliotische Verbiegung der Lenden- und unteren Brustwirbelsäule zur Folge hat. — Bezüglich des übrigen *Status* sollen nur kurz folgende Punkte berührt werden:

Kreislauforgane: o. B. Insbesondere liegen weder klinische, noch röntgenologische Zeichen einer congenitalen Herzmissbildung vor.

Respirationsorgane: o. B.

Verdauungsorgane: Ausser einigen kariösen Zähnen, die teils der ersten, teils der zweiten Dentition angehören, sind keine pathologischen Befunde zu erheben.

Nervenstatus: o. B.

Psychischer Status: o. B. Es handelt sich um ein aufgewecktes Kind von gut durchschnittlicher Intelligenz.

Körpermasse: vgl. Tab. II.

Aus der *Anamnese* sind folgende Tatsachen erwähnenswert: Das Kind stammt aus einer Bauernfamilie, in der Missbildungen, besonders Knochenanomalien, nicht bekannt sind. Die Eltern und 4 Geschwister sind gesund. Pat. wurde als viertes von 5 Kindern geboren. Die Mutter gibt an, dass sie gegen Ende der Schwangerschaft häufig Schmerzen im Mittel- und Oberbauch verspürte, was bei den 3 vorangegangenen und auch bei der folgenden Schwangerschaft nicht der Fall gewesen sei. Im übrigen sei jedoch diese vierte Gravidität, wie auch die Geburt komplikationslos verlaufen. Die Missbildung wurde sofort nach der Geburt festgestellt. Bezüglich der Entwicklung des Kindes mögen folgende

Tabelle II

Körpermasse

	Pat G. B., 10-jährig		Sollwert[1]
Körpergewicht	19.5 kg		27 kg
Gesamtkörperlänge	92.0 cm		128 cm
Scheitelsteisslänge	65.5 cm		
Kopfumfang	51.5 cm		52 cm
	Rechts	Links	
Oberarmlänge (Coracoid—Olecranon)	25 cm	24 cm	
Vorderarmlänge (Olecranon—Handgelenk)	20 cm	19 cm	
Handgelenk—Spitze des Mittelfingers	15 cm	15 cm	
Fusslänge	18 cm	19 cm	

Daten von Interesse sein: Sitzen mit 8—9 Mon., Stehen mit ca. 15 Mon., erste selbständige Gehversuche mit 18—20 Mon., Treppensteigen mit ca. 4 Jahren. Durchbruch des ersten Milchzahnes mit 7 Mon., des ersten bleibenden Zahnes mit 7 Jahren. Geistige Entwicklung und Erlernung der Sprache erfolgten in normalen Zeiten. Mit 7 Jahren trat Pat. in die Schule ein. Besonders erwähnenswert scheint uns die Tatsache, dass sie auch im Turnunterricht mitmacht. — Ernstliche Krankheiten hat das Kind bisher angeblich nicht durchgemacht. Untersuchungen wegen der bestehenden Missbildung wurden im Alter von 8, 16 und 40 Mon. vorgenommen.

Dieser Fall erregte unsere Aufmerksamkeit besonders dadurch, dass Pat. trotz der erheblich missbildeten unteren Extremitäten nicht nur stehen und gehen, sondern erstaunlich behend Treppen steigen und sogar aus 40—50 cm Höhe auf harten Boden herabspringen kann. Tägliche Märsche von 4—5 km haben keine wesentliche Ermüdung zur Folge. Schmerzen treten auch bei etwas grösseren Anstrengungen, bei längerem Stehen oder Gehen, weder in den Hüftgelenken, noch in den Beinen auf.

Bei der abnormen Kürze der Beine und dem Fehlen beweglicher Kniegelenke stellte sich uns die Frage, ob hier eine Aplasie der Ober- oder der Unterschenkel vorliege, oder ob beide rudimentär entwickelt und dabei die Knie- oder Hüftgelenke ankylosiert seien. — Zur Abklärung haben wir das Kind eingehend röntgenologisch untersucht, wobei einige interessante und selten beschriebene Knochenbefunde erhoben werden konnten.

Auf dem a.-p. *Übersichtsbild des Beckens* (Fig. 3), das den Zustand im Alter von 10 Jahren und 5 Monaten zeigt, fällt vor allem das atypische Aussehen beider Hüftgelenke auf. Gelenkpfannen sind nicht einmal andeutungsweise zu erkennen, während die »Femurköpfe», auf deren absonderliche Gestalt im Zusammenhang mit der Beschreibung des

[1] FEER: Lehrbuch der Kinderheilkunde, Jena, 1944.

Fig. 3. Becken-Übersichtsaufnahme, a.-p., im Alter von 10 Jahren und 5 Monaten.

Extremitätenskelettes noch näher eingegangen wird, nur in den Weichteilen stecken und bei liegender Pat. und freier Projektion mindestens 1 cm vom nächsten Punkt des Beckens entfernt sind. Die Foramina obturatoria erscheinen abnorm gross, die Synchondrosis ischiopubica ist beidseits auf eine Länge von 5—6 mm noch nicht verknöchert. Die Sitzbeinhöcker stehen etwas weit lateral, sind eher plump geformt und weisen eine eigenartige Knochenstruktur auf, wobei die Knochenbälkchen seitwärts und nach unten orientiert sind, als ob sie einer Zugwirkung folgten. Die bindegewebige Symphyse ist 6—7 mm breit. Die Y-Fugen sind noch grösstenteils offen. Die Beckenschaufeln entsprechen in Grösse und Form etwa dem Alter des Kindes. Ihre Knochenstruktur ist im Gebiete oberhalb der Y-Fugen eigenartig gitterförmig. Es lassen sich zudem arkadenartig angeordnete Knochenbälkchen erkennen, die von der Y-Fuge aus lateral-aufwärts ziehend den unteren Teil der Darmbeine verstärken. Das Sacrum zeigt einen relativ geringen Kalkgehalt, weist jedoch bezüglich seiner Form und seiner Verbindungen mit dem übrigen Beckenskelett keine Besonderheiten auf. Dagegen sind Steisswirbel weder auf der a.-p., noch auf der seitlichen Aufnahme nachzuweisen.

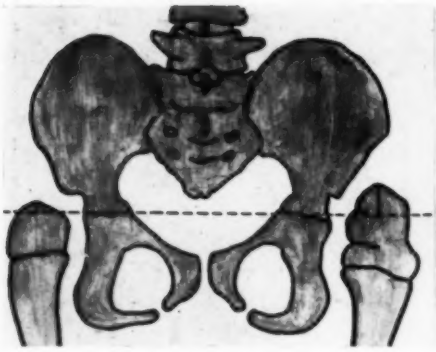

Fig. 4 a. Becken und Hüftgelenke im Liegen.

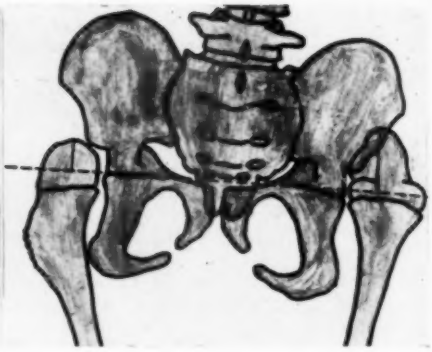

Fig. 4 b. Becken und Hüftgelenke im Stehen.

Bei dem unerwarteten Hüftgelenksbefund, der unter Berücksichtigung der grossen Foramina obturatoria mit offener Synchondrosis ischiopubica weitgehend an das Bild der congenitalen Hüftgelenksluxation erinnert, haben wir zur weitern Abklärung der Beziehungen zwischen dem Extremitäten- und dem Beckenskelett ein Beckenübersichtsbild im Stehen angefertigt. *Liegend- und Stehend-Aufnahme* sind in Fig. 4 in Form von Skizzen einander gegenübergestellt. Während auf der Liegendaufnahme (Fig. 4 a) die Distanz von der durch beide Y-Fugen ziehenen Markierungslinie senkrecht auf den höchsten Punkt des »Femurkopfes» rechts 5 mm und links 18 mm beträgt, ist sie bei der Stehendaufnahme (Fig. 4 b) rechts 18 mm und links 40 mm. Unter der Last des Körpers gleiten also beide unteren Extremitäten um ein beträchtliches Stück neben dem Becken empor. Damit erklärt sich wohl die bereits mitgeteilte Beobachtung, dass die Weichteile um beide Hüften beim Stehen weit mächtiger in Erscheinung treten als beim Liegen. — In diesem Zusammenhang ist auch ein Vergleich mit der *im Alter von 3 Jahren und 4 Mon. angefertigten Beckenaufnahme* (Fig. 5, Skizze) nicht uninteressant, standen doch damals die »Femurköpfe» inbezug auf die Y-Fugen noch bedeutend

Fig. 5. Beckenübersichtsaufnahme im Alter von 3 Jahren und 4 Monaten.

tiefer. Die Annahme, dass seither durch das tägliche Stehen und Gehen allmählich eine Verschiebung in die jetzige Lage zustande gekommen sei, ist wohl berechtigt.

Die wichtigen Ausschnitte der Röntgenbilder der *rechten unteren Extremität* sind in Fig. 6 & 7 reproduziert. Das Bein besteht aus einem einzigen langen Röhrenknochen, der proximal durch eine etwa kastaniengrosse, femurkopf-ähnliche Epiphyse begrenzt wird, während sich distal eine scheibenförmige Epiphyse findet, die einigermassen an das distale Ende einer Tibia erinnert, wobei allerdings der Malleolus internus nicht typisch ausgebildet ist. Die Epiphysen sind durch noch offene Epiphysenfugen von der Diaphyse getrennt. Der »Femurkopf» (Fig. 6) scheint im Bereiche der Epiphysenlinie lateralwärts etwas über den Schaft abgeglitten zu sein. Ein Schenkelhals ist nicht nachzuweisen. Ebenso finden sich keine Trochanterkerne. Besonders zu erwähnen sind dagegen zwei parallel verlaufende, lateral- und vorwärts orientierte, sägeartige Knochenleisten, die von der proximalen Epiphysenfuge aus ca. 6 cm weit distalwärts ziehen, und die vermutlich als Ansatzstellen von Muskeln oder Sehnen von Bedeutung sind. Die weiche Aufnahme zeigt überdies, dass sich um den rechten Femurkopf innerhalb der Weichteile zwiebelschalenartig angeordnete dichtere und hellere Zonen unterscheiden lassen, die in den erwähnten arkadenartigen Knochenbälkchen der Beckenschaufel offenbar ihre Fortsetzung finden. Letzteren dürfte somit eine statische Bedeutung zukommen, während es sich bei den zwiebelschalenartigen Weichteilverdichtungen möglicherweise um derbe Bindegewebszüge handelt, die das fehlende Hüftgelenk ersetzen.

Fig. 6. Rechtes Hüftgelenk. Hüftgelenkspfanne kaum angedeutet. »Femurkopf» im Bereich der Epiphysenlinie etwas lateral abgeglitten. Zwiebelschalenartige Weichteilverdichtungen, die sich in die arkadenartig angeordneten Knochenbälkchen des Beckenskelettes fortsetzen.

Distal artikuliert der eben beschriebene Röhrenknochen mit einem Fusswurzelknochen, der seiner Form nach als ein Verschmelzungsprodukt von Talus und Calcaneus aufgefasst werden muss. Ausser diesem ist auf

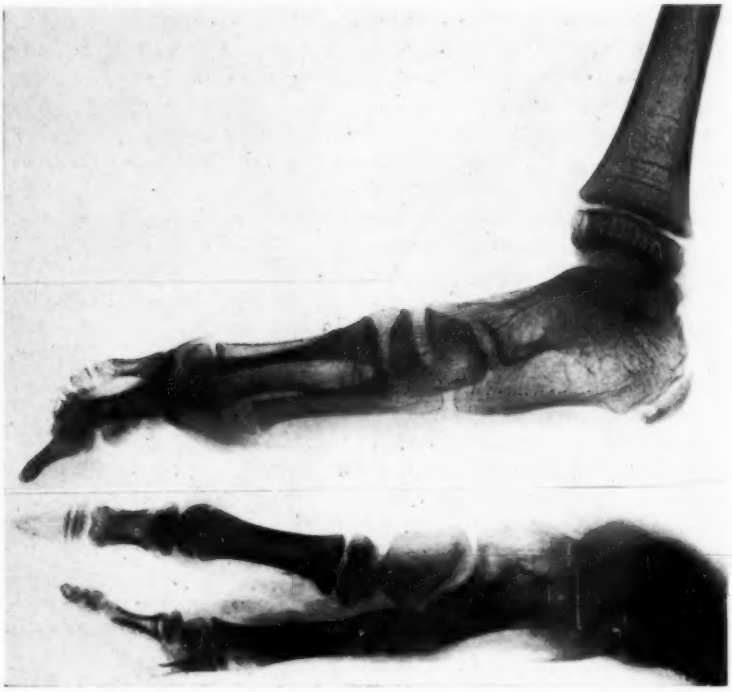

Fig. 7. Rechtes Sprunggelenk und rechter Fuss. Fehlen der Fibula, VOLKMANN'sche Sprunggelenksdeformität. Verschmelzung von Talus und Calcaneus. Aplasie einiger Fusswurzelknochen und zweier Endstrahlen. Teilweise Verwachsung der beiden lateralen Endstrahlen.

den Aufnahmen des *rechten Fusses* (Fig. 7) nur noch ein Tarsalknochen zu erkennen, der wohl dem Cuneiforme I entspricht, sofern es sich nicht um ein Kondensationsprodukt mehrerer Tarsalia handelt. An das rudimentäre Fusswurzelskelett schliessen sich 3 Metatarsalia an. Die proximalen Enden der beiden lateralen sind mit einander verwachsen, der mediale dagegen ist frei und zeigt auch sonst keinerlei Besonderheiten. Während die normal geformte Grosszehe zweigliedrig ist, setzen sich die beiden lateralen Zehen aus je 3 normal gestalteten Phalangen zusammen.

Die *linke untere Extremität* besteht ebenfalls nur aus einem einzigen langen Röhrenknochen und einem rudimentär entwickelten Fuss, die mit einander in gelenkiger Verbindung stehen. Das proximale Ende (Fig. 3) wird durch einen etwa kastaniengrossen »Femurkopf« gebildet, der im Gegensatz zur rechten Seite durch Vermittlung einer etwa 1 cm hohen,

etwas breiteren Knochenscheibe mit dem Schaft verbunden ist, während sich am distalen Ende eine ungefähr 1 cm hohe Epiphyse erkennen lässt, deren Gelenkfläche eine gewisse Ähnlichkeit mit derjenigen des distalen Tibiaendes zeigt. Zwischen dem »Femurkopf» und der erwähnten Knochenscheibe, sowie zwischen dieser und dem Schaft und schliesslich zwischen dem Schaft und der distalen Epiphyse finden sich noch grösstenteils offene Epiphysenfugen.

Die Aufnahme des *linken Fusses* (Fig. 8, Skizze) zeigt ein ähnliches Bild wie rechts. Es besteht auch hier eine Verschmelzung des Talus und

Fig. 8. Linker Fuss. (Skizze nach dem Röntgenbild.) Verschmelzung von Talus und Calcaneus, Aplasie einiger Fusswurzelknochen und zweier Endstrahlen. Keine Verwachsung der bestehenden lateralen Metatarsalia.

Calcaneus. Daneben lassen sich noch zwei weitere Fusswurzelknochen, wahrscheinlich die Cuneiformia I und II nachweisen. Die 3 Metatarsalia und die 3 Zehen zeigen keine Besonderheiten.

Über die Resultate der *übrigen röntgenologischen Skelettuntersuchung* berichten wir nur ganz kurz. Weitere Fehlbildungen sind nicht vorhanden. Erwähnt sei aber, dass zwischen dem rechten und linken Handgelenk eine geringfügige Entwicklungsdifferenz besteht, indem links das Pisiforme bereits als gut erbsgrosses Knöchelchen zu sehen ist, während es rechts noch vollständig fehlt. — Schädel, Wirbelsäule, Thorax, Schultern, Ellbogen und Hände entsprechen in Form und Entwicklungsgrad der Norm.

Zusammenfassend lassen sich die *Ergebnisse der Röntgenuntersuchung* folgendermassen formulieren:

1. Atypisches Verhalten beider Hüftgelenke, wobei nebst der Formanomalie der Extremitäten eine doppelseitige kongenitale Hüftgelenksluxation vorliegt. Besondere Beachtung verdient dabei die Tatsache, dass diese Hüftgelenke trotzdem weitgehend funktionstüchtig sind, was vermutlich durch die Bindegewebsverstärkungen in ihrer Umgebung, die wir auf einer weichen Aufnahme erkennen können, bedingt ist.

2. Fehlen der Steisswirbel.

3. Symmetrisches Fehlen zweier langer Röhrenknochen der unteren Extremitäten, wobei nicht sicher zu entscheiden ist, ob es sich um Tibia

und Fibula, oder um Femur und Fibula handelt. Möglicherweise sind die als »Femurkopf» angesprochenen Knochen die sehr rudimentären Reste der Femora, während Schaft und distale Epiphyse der Tibia entsprechen.

4. Fehlen der Patella.

5. Symmetrische Verschmelzung von Talus und Calcaneus.

6. Aplasie oder Verschmelzung weiterer Fusswurzelknochen, mit Aplasie zweier Endstrahlen.

7. Geringe Entwicklungsdifferenz zwischen dem rechten und linken Handgelenk.

Sämtliche Fehlbildungen erreichen rechts einen etwas höheren Grad als links; so ist links zwischen »Femurkopf» und Schaft eine Knochenscheibe eingelagert, die rechts völlig fehlt; weiterhin können links total 3 Fusswurzelknochen isoliert nachgewiesen werden, rechts nur deren zwei, und schliesslich sind links die lateralen Metatarsalia völlig getrennt, während sie rechts im proximalen Viertel eine partielle Verschmelzung zeigen. Es besteht somit links eine etwas stärkere Tendenz zur Norm, die sich auch am Handskelett durch die bereits vorhandene Anlage des Pisiforme manifestiert. — Die Feinstruktur der Knochen ist überall, selbst bei den schwer deformierten Knochen, in normaler Weise ausgebildet, es sind keinerlei Zeichen degenerativer Knochenprozesse zu finden.

Die *Ätiologie* solcher Missbildungen ist einstweilen noch unklar. Die früher angenommene, amniogene Abschnürung der Extremitäten kann bei symmetrischer Fehlbildung mit hoher Sicherheit abgelehnt werden. Der hier mitgeteilte Fall dürfte dafür besonders beweisend sein, sind doch bei fehlender Tibia (oder Femur) und Fibula die Füsse, wenn auch rudimentär, vorhanden. — Nach SCHINZ (5) ist in einzelnen Fällen Vererbung einwandfrei festzustellen. Meistens ist jedoch ein erbliches Moment schwer zu beweisen, da diese stark missbildeten Individuen entweder früh sterben oder wenigstens nicht zur Fortpflanzung kommen.

Wie eingangs erwähnt, haben wir auch bei unserem Fall keinen Anhaltspunkt für Vererbung. Wir müssen vielmehr annehmen, dass es sich bei diesen Aplasien um eine sprunghaft aufgetretene Verlustvariante handelt, die möglicherweise recessiv vererbbar ist und nur bei Homozygotie in Erscheinung tritt, während sich bei Heterozygotie höchstens leichte Störungen, z. B. Fehlbildungen einzelner Finger oder Zehen zeigen. Für diese Möglichkeit spricht z. B. auch ein von PATEL-CHARTON (4) mitgeteilter Fall. Dabei handelt es sich um einen 34-jährigen Mann, dessen Kopf, Hals und Rumpf anatomisch und funktionell keine Besonderheiten aufwiesen, während die unteren Extremitäten ähnlich schwere Deformitäten zeigten, wie bei unserer Patientin. Darüber hinaus waren aber auch die Arme hochgradig missbildet. Dieser Mann war mit einer

normal entwickelten Frau verheiratet und Vater eines durchaus normal gewachsenen Sohnes.

Bei Durchsicht der Literatur haben wir sonst einzig in einer Arbeit von KRICHLER (2) ein ähnliches Beispiel (sein Fall 9) einer doppelseitigen Aplasie zweier langer Röhrenknochen der untern Extremitäten gefunden. Bei diesem Fall liessen sich jedoch im proximalen Abschnitt der Extremitäten (Bereich der »Hüftgelenke») röntgenologisch etwas ausgedehntere Knochenformationen, vor allem noch ein deutlicher Femurrest erkennen. Diese Missbildung ist in den Rahmen einer teratologischen Reihe gestellt und als beidseitige Femur-Aplasie — mit Fibula-Aplasie und Endstrahldefekten als Begleiterscheinungen — interpretiert. Die ganze Beobachtungsreihe KRICHLER's, angefangen bei geringen Störungen des Längenwachstums der Fibula bis zu den schwersten Aplasien langer Röhrenknochen, wird als Bestätigung der GEGENBAUR'schen Theorie aufgefasst, wonach sich der Extremitätenstrahl (Archipterygium der Säugetiere) an der unteren Extremität vom Femur über die Fibula in den lateralen Fussrand zur kleinen Zehe erstreckt.

In seiner Arbeit über den kongenitalen Femurdefekt berichtete auch NILSONNE (3) über prinzipiell ähnliche, wenn auch geringgradigere Defekte von Femur, Fibula und lateralen Endstrahlen.

K. H. BAUER (1) nimmt nun zur Erklärung der Genese derartiger Hypo- und Aplasien von Skelettelementen allgemein die Mutation eines einzigen Gens an, das die Disposition für Knochenmissbildungen schaffe. Es scheint uns durchaus denkbar, dass ein solcher Gendefekt einmal der tiefere Grund für die Aplasie des GEGENBAUR'schen Extremitätenstrahles darstellen kann. Im Lichte dieser Theorie dürfte unser Fall als doppelseitige, vollständige Aplasie des GEGENBAUR'schen Extremitätenstrahles bei intakter Tibia und intakter medialer Fusseite interpretiert werden, was mit unsern Röntgenbefunden recht gut übereinstimmt.

ZUSAMMENFASSUNG

Bericht über ein 10-jähriges Mädchen, das eine beidseitige Aplasie zweier langer Röhrenknochen der unteren Extremitäten und zahlreicher Fussknochen aufweist. Besonderes Interesse verdient der Fall deshalb, weil das Kind im übrigen normal entwickelt ist und trotz seiner Missbildungen eine erstaunlich gute Bewegungsfähigkeit erworben hat. Die ätiologischen Möglichkeiten solcher Missbildungen werden diskutiert.

SUMMARY

Report of a case of a girl, aged 10, who suffered from bilateral aplasia of two bones of the lower extremities and numerous bones of the feet. It is interesting to note that the patient was otherwise normally developed and able to walk with remarkable ease. The etiological factors concerned in these malformations are discussed.

RÉSUMÉ

L'auteur relate le cas d'une fillette de 10 ans qui présentait une aplasie bilatérale de deux des grands os des jambes et de plusieurs petits os des pieds. Le cas est spécialement intéressant par le fait que l'enfant s'est normalement développée pour le reste et a acquis une mobilité étonnante malgré sa difformité. L'étiologie de ces malformations est discutée.

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FROM THE ROENTGENOLOGICAL RESEARCH LABORATORY, KAROLINSKA SJUKHUSET, STOCKHOLM, SWEDEN

ELECTROKYMOGRAPHY OF THE PULMONARY ARTERY IN CASES OF PULMONARY STENOSIS[1]

by

Sven Roland Kjellberg and *Ulf Rudhe*

Pulmonary stenosis — often associated with patent interauricular septum — seems to occur more frequently, and consequently represents a more important clinical condition, than was previously supposed. Thus, during the last years a rapidly growing series of cases has been reported in the literature (CURRENS, KINNEY and WHITE, SELZER et al., GREENE et al., POLLACK et al., ALLANBY and CAMPBELL, CAMPBELL and HILLS, HOLLING and ZAK, HANSEN et al., MANNHEIMER et al. amongst others). An exact interpretation was in most cases due to the aid of cardiac catheterization and angiocardiography.

Reports of the use of *electrokymography* as a diagnostic method in congenital heart disease are comparatively few. The experience gained in this field seems to be only fragmentary. However, a fairly extensive study of the pulsations in the lung fields in different forms of congenital heart disease has been made by JORGENS et al. In a series of 12 cases of pulmonary stenosis, associated with various anomalies, they found a reduced height of the systolic upward deflections in the electrokymographic records. As far as we know a strictly reliable method of calibrating the electrokymograph has not been evolved. The interpretation of these findings may thus be difficult.

In an examination of 11 cases of coarctation of the aorta from an electrokymographic point of view, KJELLBERG and RUDHE showed the presence of typically altered records of the contour movements of the descending aorta. These records were characterized by a more or less sine-curve appearance, as well as by a smoothing out of the incisura, the dicrotic wave and the small secondary deflections. This was in marked contrast to the normal electrokymograms. The abnormal pattern of the

[1] Submitted for publication, April 26, 1951.

tracing was correlated to the deformation of the pulse wave registered distal to the stenosis in such cases.

The normal electrokymographic tracings from the descending aorta and the pulmonary artery are in many respects alike. The volume changes of the vessels more or less parallel the pressure changes of the pulse waves. Only to a small extent do the positional movements of the artery seem to influence the fundamental form of the records. It is reasonable to assume that the circulatory mechanism behind the stenosis may principally be the same, whether the latter is located in the descending aorta, pulmonary valves or infundibular region. Moreover, from an anatomical point of view, a poststenotic dilatation is a frequent finding in coarctation of the aorta as well as in cases of pulmonary stenosis. Hence, it was supposed that an abnormal electrokymogram of the pulmonary artery might be found even in the latter type of malformation.

Eight such cases have been examined in one year at the Karolinska sjukhuset. They are verified by cardiac catheterization. The diagnostic aspect of electrokymography in three of these cases will be illustrated.

The electrokymograph used was the same as in previous investigations (1949, 1950). The speed of the film was 100 mm per second. There was simultaneous recording of the Ecg (Lead II) and in cases 2 and 3 of the phonocardiogram as well. In the latter case the pressure curve of the pulmonary artery was recorded at the same time during cardiac catheterization. The slit of the photocell was placed over the border of the shadow of the main pulmonary artery and the examination was made both in the sitting and recumbent position. The lag of the recording was 0.01—0.02 sec.

Case Reports

Case 1. K. Å., female sixteen years old. Referred to the Hospital on account of occasionally easily becoming tired and a coarse systolic murmur heard over the whole precordium. The maximal intensity of the murmur was heard over the pulmonic area, where the second sound was faint. No dyspnoea at rest. Moderate cyanosis increasing during exercise.

Fluoroscopy and *roentgenography* showed a moderately enlarged heart. Heart volume in prone position 670 ml. The right ventricle was responsible chiefly for the enlargement. Moderate enlargement of the main pulmonary artery and a slight concavity below this shadow in the pulmonary conus region. There was a decrease in the peripheral vascular markings in the lung fields.

Electrokymography showed an abnormal pulmonary artery tracing characterized by a slowly rising upward deflection and reduction of the incisura and secondary waves. A tentative diagnosis of pulmonary stenosis was made.[1] Fig. 1. According to information kindly supplied by Prof. CRAFOORD after heart catheterization this diagnosis was later confirmed.

[1] This case has previously been presented by one of us (U. R.) at the meeting of Svenska Föreningen för Medicinsk Radiologi, October 7, 1950.

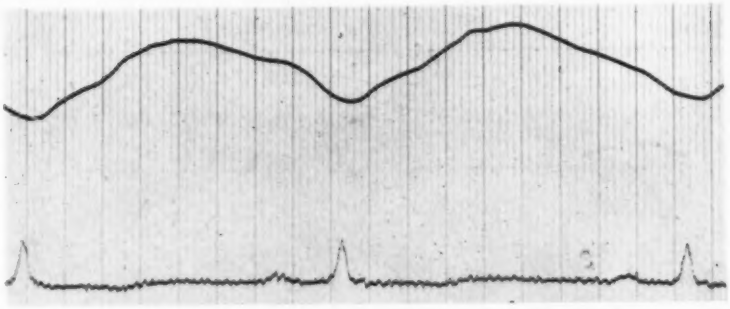

Fig. 1. Electrokymogram of the pulmonary artery in case 1.

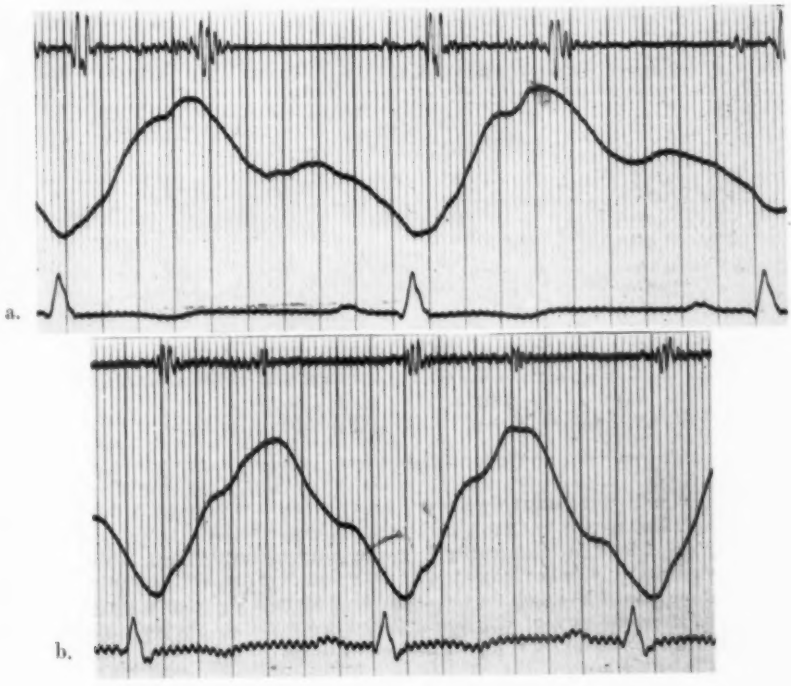

Fig. 2. Electrokymogram of the pulmonary artery in case 2.
a. recumbent position. b. sitting position.

Case 2. K. T., male 18 years old. Congenital heart disease of unknown nature discovered in infancy. Always dyspnoeic and tired easily while running. Marked tachycardia on exertion. Slight dyspnoea at rest.

Loud coarse systolic murmur of long duration, with maximal intensity over the third left interspace; audible also at the back. Blood pressure: 100/75.

Fig. 3. Electrokymogram of the pulmonary artery in case 3.
a. During heart catheterization (recumbent position) with simultaneous recording of the pressure curve.
1. Pressure curve. 2. Electrokymogram.
b. Sitting position

Fluoroscopy and *roentgenography* showed an enlarged heart. Heart volume in prone position 800 ml. Enlargement of the right ventricle. Considerable dilatation of the main pulmonary artery. Slight increase in the vascular markings in the lung fields.

Electrokymography: Slowly rising upward deflection with an anacrotic wave. Absence of the normal incisura. The notch on the descending limb of the tracings, appears about 0.10 of a sec. after the end of the second sound in the recumbent position and 0.14 in the sitting position in spite of a higher heart-rate in this posture. It may be caused by positional shifts of the artery. Fig. 2.

Heart catheterization

	Pressure		Oxygen content
Right auricle	15/3 mm Hg	Vena cava	11.1 vol. %
Right ventricle	65/5 » »	Right auricle	11.1 » »
Pulm. artery	15/0 » »	Right ventricle	11.1 » »
		Pulm. artery	10.9 » »

Arterio-venous O_2 diff. 6.2 vol %.

D. Pulmonary stenosis.

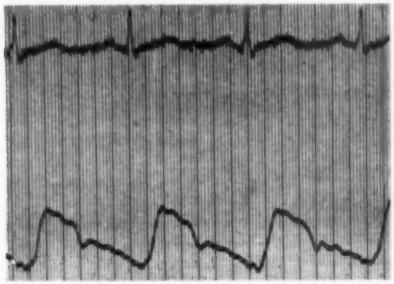

Fig. 4. Pressure curve recorded during heart catheterization of a normal case.[1]

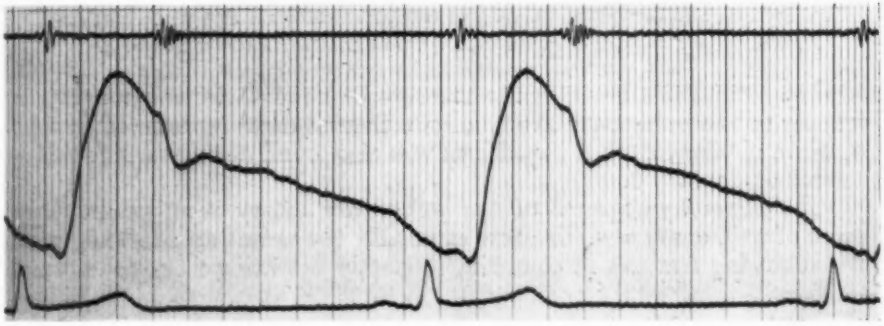

Fig. 5. Electrokymogram of the pulmonary artery of a normal case.

Case 3. K. H. female thirty-seven years old. For 5 years her activities had been limited because of increasing complaints of dyspnoea even on slight exertion.

No dyspnoea at rest. Suggestion of cyanosis of the lips.

Faint systolic murmur best heard over the third left interspace. Somewhat accentuated pulmonic second sound. Blood pressure: 115/95.

Fluoroscopy and roentgenography showed an enlargement of the heart. Heart volume in prone position 760 ml. Enlargement of the right ventricle and possibly slight dilatation of the left ventricle.

Considerable dilatation of the main pulmonary artery and moderate enlargement of its left and right branch. No abnormal vascular markings in the peripheral lung fields.

Catheterization of the heart:

	Pressure		Oxygen content
Right auricle	18/5 mm Hg	Vena cava	8.8 vol. %
Right ventricle	125/15 » »	Right auricle	9.8 » »
Pulm. artery	30/5 » »	Right ventricle	10.7 » »
		Pulm. artery	3.5 » »
		Left pulm. artery	3.9 » »

[1] The curve is reproduced by courtesy of Dr. S. R. JOHNSON.

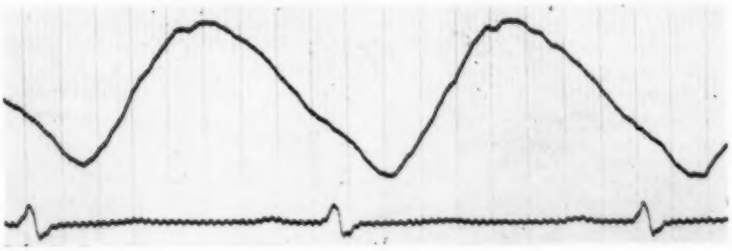

Fig. 6. Electrokymogram from the descending aorta distal to the stenosis in a case of coarctation of the aorta.

Electrokymography: Slowly rising upward deflection and nearly complete absence of the incisura and secondary waves. The tracing resembles a sine-curve, fig. 3.

D. Pulmonary stenosis + septal defect (interauricular? interventricular?).

The similarities between the pressure curve and electrokymographic tracing of the pulmonary artery of two normal cases reproduced in figs. 4, 5 are obvious. Such a parallelism was also found during cardiac catheterization in case 3.

The electrokymograms of the pulmonary artery recorded in three cases of pulmonary stenosis show essentially the same pattern. They have the following features in common, distinctly deviating from the normal findings.

1. More or less sine-curve appearance with slowly rising upward deflection.

2. Partial reduction or complete absence of the incisura and the secondary waves of the descending slope.

This pattern of the tracings was never observed in a large series of normal and pathological cases other than in pulmonary stenosis. The changes are prominent. They are compatible with the altered records from the descending aorta in isthmic stenosis fig. 6. Looked upon as an electrokymographic entity, they may form the basis of a special diagnosis of pulmonary stenosis.

As this malformation of the heart is sometimes amenable to successful surgical correction by valvulotomy (BROCK), there are increasing demands for accurate diagnosis. Electrokymography affords a simple method of recognizing the presence of such a lesion, the exact nature of which must be further evaluated by cardiac catheterization and angiocardiography.

Three of the cases have kindly been placed at our disposal by GUNNAR EKSTRÖM, M. D. and one by GUNNAR MALMSTRÖM, M. D., for which we wish to express our appreciation.

SUMMARY

The diagnostic significance of the electrokymographic tracings of the pulmonary artery in 8 cases of pulmonary stenosis is stressed. The records are analogous to those described for the descending aorta in cases of coarctation of the aorta.

ZUSAMMENFASSUNG

Die diagnostische Bedeutung der elektrokymographischen Darstellung der Pulmonalarterie in 8 Fällen von Pulmonalstenose wird betont. Die Kurvenbilder sind analog jenen, welche in Fällen von Isthmusstenose beschrieben wurden.

RÉSUMÉ

Description du tracé obtenu à l'aide du tambour électrique dans 8 cas de rétrécissement de l'artère pulmonaire. L'image coïncide avec celle obtenue au niveau de l'aorte descendante dans les cas de rétrécissement de l'isthme.

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IMPROVEMENTS OF THE EQUIPMENT USED IN THE MASKING METHOD OF RADIOGRAPHIC PRINTING[1]

by

Ove Mattsson

The method for printing radiographs with contrast levelling described by the author (1951) has its chief advantage in its great simplicity and in the fact that no special apparatus is required apart from that available in an ordinary photographic laboratory or radiographic clinic. The results are also similar whether simple tools — as described in the above paper — or the arrangements described below are used.

The author's method consists in control of the illumination of the radiograph under printing by the use of a wet print of it on thin document paper as a translucent mask. This is placed close to the radiograph and in register with it and transmits varying amounts of light. Thus a selective control is obtained; the densest areas of the radiograph will be most intensely and the most transparent areas least intensely illuminated. The effect is a levelling of contrast with an extreme improvement in the reproduction of details within areas of widely different densities on the same image.

However, it has proved desirable to design an apparatus for large-scale printing of radiographs, the handling of which should be as simple and rapid as possible, while utilizing the method to the utmost. The apparatus described below is the outcome of a series of experiments (LAURIN and MATTSSON) and has proved ideal for its purpose.

It consists of the largest model available in the market of a standard copying apparatus — ordinarily used for reflex copying — which after the addition of some special technical arrangements[2] proved extremely suitable for its purpose. The illumination is provided by fluorescent tubes and incandescent bulbs, and four different levels are available, two for darkroom work and two for exposure. This facilitates the handling, as the document paper and the regular printing paper have different de-

[1] Submitted for publication, May 16, 1951.

[2] The construction was performed by Molander & Son, Stockholm.

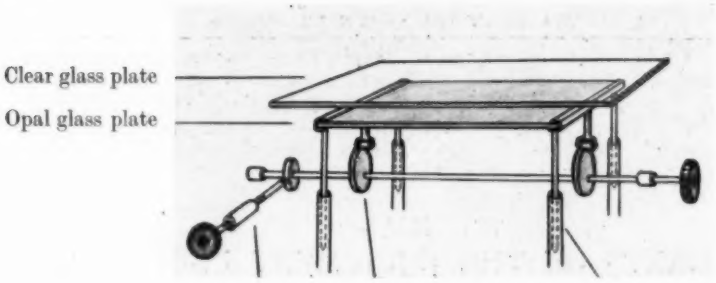

Fig. 1. Construction of the movable opal glass frame.

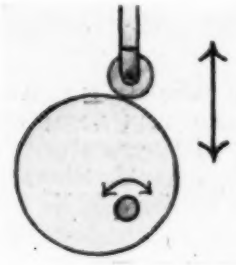

Fig. 2. Detail of eccentric mechanism.

grees of sensitivity. A timer controls the exact length of the exposure in every case. Film and paper are held flat by air pressure acting on a thin rubber pad, which eliminates the use of weights. The principle is similar to that of the blood pressure cuff. The hinged cover which houses the pressure-pad arrangement, is laid down and locked in position and air pressure is applied by a simple handpump and the size of the pressure is read on a gauge. The distance between the masking print and the radiograph is varied by using a movable opal-glass frame for the wet mask; this frame may be raised or lowered at will and is placed below the clear glass plate on which the radiograph is laid.

A hand-wheel is provided for controlling the position of the frame, another for locking the mechanism in position. The principal components of the construction are eccentrics and parallel motion guides in the shape of telescopic tubes. The opal glass plate may be pulled out, sliding in the frame like a drawer, which facilitates the application of the wet mask on the glass plate. Celluloid plates of appropriate size and thickness protect the radiographic film from the wet paper in printing.

Fig. 1 shows the elements of the construction. Beneath the clear glass plate is the opal glass frame, the height of which can be controlled by

Fig. 3. Radiograph (a) lying on the clear glass plate. Masking print (b) on the opal glass plate, which is pulled out.

Fig. 4. The mask has been brought into register with the radiograph. In contact with this an unexposed printing pater. The cover is being lowered to prepare the apparatus for exposure.

hand-wheels acting on the eccentric mechanism, a detail of which is illustrated in fig. 2.

Fig. 3 is a view of the apparatus being prepared for exposure; a is the

radiograph, b is the masking print on the opal glass plate, which is pulled out. The black flat surface on the inside of the cover is the rubber pad, which is not yet in action. On the front side of the apparatus are the timer and levers for controlling air pressure and height of opal glass frame. On fig. 4 the masking print has been brought into register, a printing paper is placed in contact with the radiograph and the cover is being lowered.

SUMMARY

The author describes an arrangement for rapid printing on a large scale of radiographs according to his earlier described masking method of contrast levelling.

ZUSAMMENFASSUNG

Der Verfasser beschreibt eine Vorrichtung zum Schnellen Kopieren in grosser Zahl von Röntgenbildern, entsprechend seiner früher beschriebenen Methode zum Kontrast-Ausgleich.

RÉSUMÉ

L'auteur décrit un appareil permettant l'impression rapide et à grand tirage des radiographies selon sa méthode précédemment décrite pour atténuer le contraste par filtrage.

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POLYOSTOTIC FIBROUS DYSPLASIA OF BONE[1]

by

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The term »osteitis fibrosa» appears to be disappearing from the clinical and roentgenological terminology. In 1926 hyperparathyroidism was separated from the group of »osteitis fibrosa» diseases, and in 1937 followed the separation of polyostotic fibrous dysplasia of bone as suggested by ALBRIGHT and collaborators (1). UEHLINGER (2) has subsequently given an account of the identical disease under the designation osteofibrosis deformans juvenilis. The name introduced in 1938 by LICHTENSTEIN (3), however, has gained general support in the past decade.

According to LICHTENSTEIN & JAFFE (4) more than 90 cases of this disease were already known in 1942, and numerous cases have been reported later, particularly in American literature, and in European quarters. ALBRIGHT and co-workers in their paper of 1937 made it clear, that in females skeletal manifestations of the type polyostotic fibrous dysplasia were not infrequently concurrent with precocious puberty and anomalous pigmentation of the skin, a sign triad which is now generally called Albright's syndrome. The combination of »osteitis fibrosa» in disseminated form with precocious puberty was previously known, and has been described amongst others by KIENBÖCK (5) in 1933.

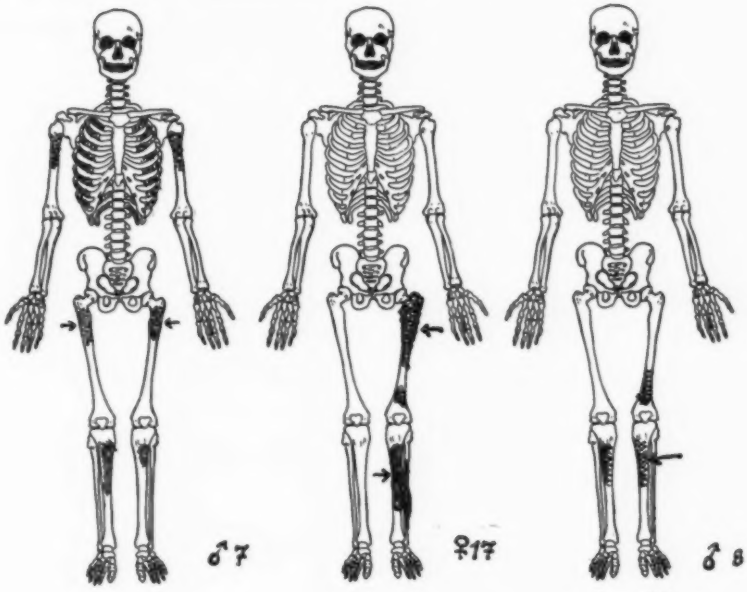
Hitherto Albright's syndrome has been observed exclusively in females. The typical skeletal changes occur, however, also in females far more frequently as the only sign and without the other two signs of the triad. It is probably also for this reason that LICHTENSTEIN & JAFFE regard the skeletal lesion as the central point in the syndrome, and maintain that the other two signs occur only when the lesion shows extensive spread in the skeleton. There appears to be exceptions from this rule, however.

[1] Submitted for publication, Febr. 3, 1951.

The causation and the nature of the disease seem to be unknown. ALBRIGHT has suggested that the peculiar spread of the disease, with involvement of the osseous tissue, skin and endocrinous organs, points to a lesion of the central nervous system, possibly in the region of the hypothalamus. UEHLINGER is of the opinion that the causal relation must be looked for in disturbances of the metabolism of the endocrinous organs, more especially that of the suprarenal glands. LICHTENSTEIN & JAFFE claim that the cause of the skeletal lesion is a congenital anomaly of the mesenchyma. Even though the lesion invariably starts in childhood, it is difficult to understand how it is possible to uphold this theory, seeing that polyostotic fibrous dysplasia — judged by all available information — originates in cartilaginous pre-formed bones, or connective tissue pre-formed bones, with normally developed cortex, and which has therefore most probably had a normal pre-morbid development. There are no data available to prove that the lesion is hereditary.

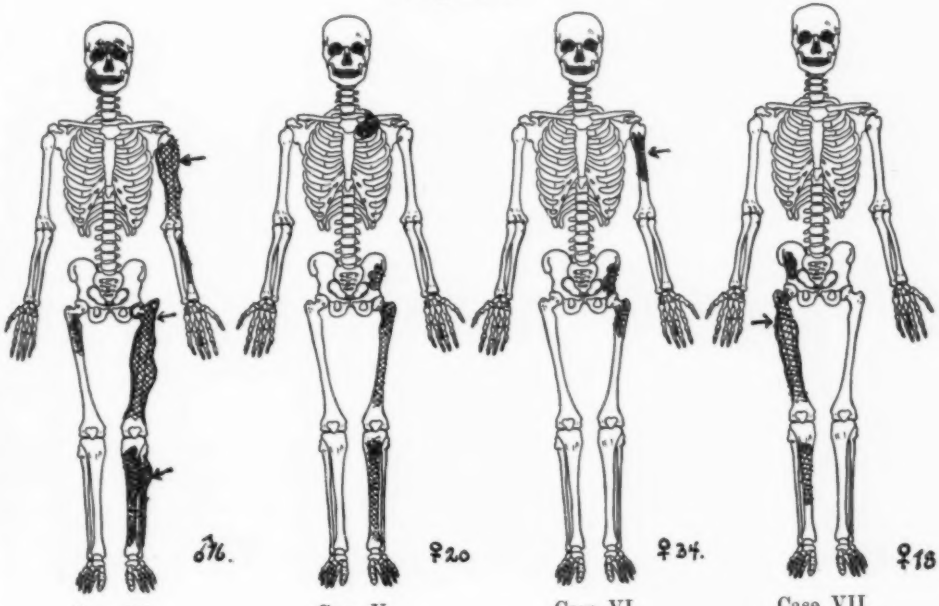
At the only complete post-mortem examination hitherto of a patient with Albright's syndrome STENBERGH & JONES (6) found a marked hypertrophy of the basophil apparatus of the hypophysis with formation of microscopic adenomas. Furthermore, atrophy of the suprarenal gland cortex was demonstrated, and in contrast a relative hypertrophy of suprarenal gland marrow tissue. The thyroid gland was somewhat hyperplastic. There were no demonstrable lesions in the central nervous system.

So far the biochemistry of bone has been so little elucidated, that it is difficult to form any definite idea of the biochemical basis of the skeletal lesion in polyostotic fibrous dysplasia. It is natural to suppose that the demonstrated hyperplasia of the basophil apparatus of the hypophysis with consequently increased formation of adrenocorticotropic hormone, is secondary, as compared with the atrophy of the suprarenal gland cortex, as an attempt at increasing the production of corticosterones, particularly 11-oxycorticosterone. The hypofunction of the suprarenal gland cortex may possibly explain the anomalous pigmentation, but hardly the increased production of cortico-N-hormone on which depends the development of the pubes and the axillary hair. It will not explain the early menarche or the peculiarly segmental localization of the disease in the skeletal system, which latter circumstance indicates that local factors play a certain rôle. The mobilization of ionized phosphorus from organic or inorganic phosphates by phosphorylases or phosphatases, is correlated with the corticosterones. The closer mechanism, however, is unknown. It is supposed that the corticosterones catalyze the phosphorylation. It has not been possible to demonstrate appreciable pathological values of alcaline monophosphoresterase in serum from individuals suffering from polyostotic fibrous dysplasia. Values slightly exceeding the normal are not infrequently found in children and



Case I. Case II. Case III.

Diagram I.



Case IV. Case V. Case VI. Case VII.

Diagram II.

Fig. 1. Fig. 2 a. Fig. 2 b. Fig. 2 c.

adolescents. Examinations as to phosphatase activity in bone removed by biopsy have never been known to have been carried out in individuals suffering from fibrous dysplastic skeletal lesions.

The disease must be said to be rare. In the course of the past decade seven cases of polyostotic fibrous dysplasia, 4 females and 3 males, have been observed in the Oslo University Hospital, Department of Roentgenology. None of these cases have been complicated by precocious puberty or anomalous pigmentation of the skin. The localization of the disease in individual cases, the sex, and age of the patient at the time of discovery of the disease, together with possible fractures and deformations, will be seen from the diagrams on page 147.

Polyostotic fibrous dysplasia commences in childhood. It is generally stated that the lesion subsides in the years following puberty, though it is not always the case. The site of predilection of the lesion in the skeleton is the long bones, most frequently the femur and the tibia. The lesion may be of monomelic or bimelic type. The unilateral, bimelic localization appears to be the most usual. Where the lesion is most pronounced it is generally bilateral and polyostotic. The most frequent

Fig. 3. Fig. 4.

localization of the lesion after the long bones is ribs and bones of the skull, less frequently the pelvic bones. The vertebrae are rarely involved.

In a long bone the fibrous dysplastic process is localized to the metaphyses and the diaphyses. The epiphyses are never attacked primarily, but may be involved in the process when they are ossified in cases where the lesion continues to be active after growth has been completed (fig. 1). The process apparently starts centrally in the bone, from the marrow space, and gradually hollows out the cortex from the inside. On this hollowing of the cortex depends one of the most essential roentgendiagnostic features. The skeletal region involved becomes more or less translucent, and the resemblance to a cyst may be marked in the initial stage. The translucent region, however, appears to become covered at an early stage by a diffuse veiling, due to the formation of numerous fine bone spicules in the tissue, which replace the destroyed osseous tissue. These spicules consist of immature, primitive osseous tissue without functional organization, and rarely reach a size to enable them to give individual markings on the roentgenograms. Thus one gets the »ground glass» appearance or »smudging» as it is also called, another important roentgenological criterion. The increasing mass of reparatory primitive osseous tissue will gradually give a roentgenological impression of sclerosis, particularly in the bones of the skull. However, normal osseous tissue never develops and the bones involved will present characteristic roentgenological changes throughout life.

Fig. 5. Fig. 6 a. Fig. 6 b.

The roentgen diagnosis of polyostotic fibrous dysplasia is generally simple. The diagnosis is established on the basis of the peculiar localization of the lesion in the skeleton, the typical scallopings of the cortex of the long bones from within and the »ground glass» appearance. The diagnosis is also supported by the relatively sharply defined border of the lesion against adjoining skeletal tissue. As a rule portions only of a bone are involved. There is no periosteal reaction and no subperiosteal formation of cysts.

The special features of the roentgenological picture appear from fig. 2. When the process inside a bone has only just commenced the roentgenological picture is not very typical. Fig. 3 shows a fibrous dysplastic process at an early stage (case 1). The small translucent area with the sclerotic margin in the proximal humeral metaphysis permits no accurate roentgenological diagnosis. In fig. 4 — one and a half year later — the excavation of the cortex from the marrow space is already distinct, and a »ground glass» appearance has developed. If the lesion is extensive deformations are frequent. In the femur the development of »shepherd's crook» deformation (fig. 5) is not infrequent if the lesion involves the femoral neck. Just as frequently the lesion seems to lead to more or less

pronounced thickening of the bones (fig. 6). This specially applies to the bones which have met with several fractures. Often it is these that lead to the discovery of the lesion. One only of the seven cases in this series has had no fractures. The fractures heal practically speaking in normal time, and thus there appears to be no fault with the periosteal formation of new bone. Re-fractures are frequent.

Fig. 7.

The lesion gives no significant changes to the content in the blood serum of calcium, phosphorus or alcaline monophosphoresterase. In none of the cases reported here we found appreciable pathological values on repeated examinations.

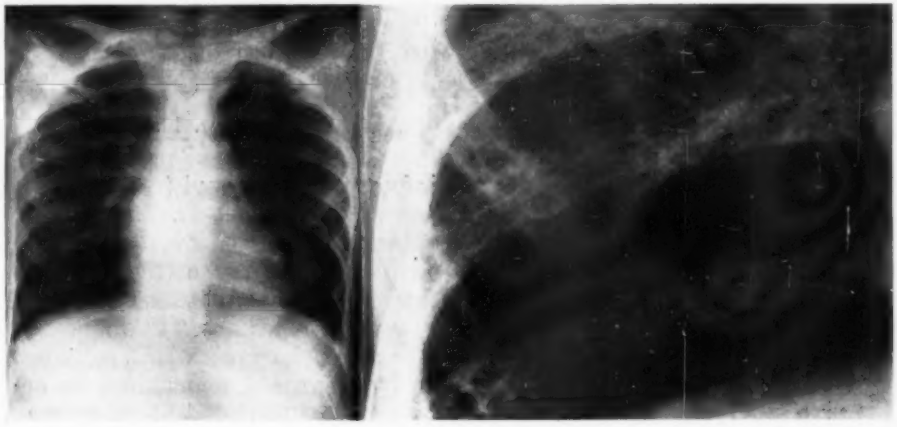


Fig. 8.

The bones of the skull were involved in one case only. In two cases there was rib involvement. In case 5 the first left rib was considerably thickened and deformed. The roentgenogram (fig. 7) presented appearances which were reminiscent of a chondroma. In case 1 the rib involvement was of a totally different type. The changes in the long bones in this case also leave no doubt as to the diagnosis. The roentgenological appearances presented by the rib involvement, however, suggest xanthomatosis (Hand-Schüller-Christian's disease — multiple myelomas) (fig. 8).

In the osseous regions affected by polyostotic fibrous dysplasia the histological picture is dominated by fibrous connective tissue. Small

Fig. 9.

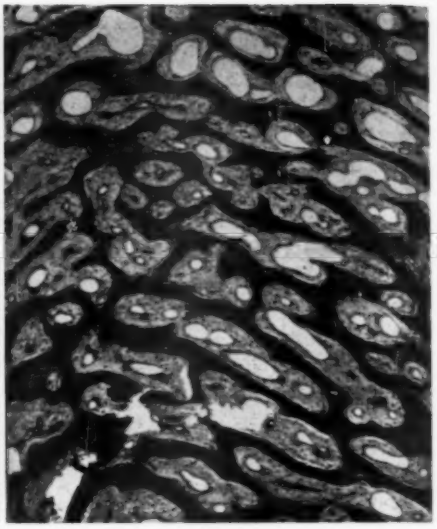


Fig. 10.

areas of cartilaginous tissue are not infrequently observed (fig. 9). Foam cells have been observed, though never in great numbers. There are as a rule few osteoblasts. Numerous trabeculae of immature, non-lamellar osseous tissue develop in the fibrous connective tissue (fig. 10). The connective tissue demonstrates a varying degree of vascularization and varying content of cells. The mass of primitive osseous tissue increases with the age of the patient, especially after the process has subsided. Roentgenologically this is expressed by increased »smudging» to approximated sclerosis (fig. 1). There is never development of normal, mature, functionally fully valid and architectonically organized bone in the osseous regions involved. DOCKERTY and collaborators (7) emphasize that the histopathological picture of polyostotic fibrous dysplasia is never so typical as to make a differential diagnosis between hyperparathyroidism and polyostotic fibrous dysplasia on it alone possible. The differential diagnosis between these two conditions, however, is easily established by the roentgenograms, and the demonstration of the characteristic serum changes of hyperparathyroidism is decisive in doubtful cases.

ALBRIGHT (8) has refuted attacks from SNAPPER (9) and THANNHAUSER (10) on the justification of upholding polyostotic fibrous dysplasia as a nosological entity. Polyostotic fibrous dysplasia is not an expression of lipoid granulomatosis (xanthomatosis), as the cholesterol

values in the blood serum are normal. Only rarely have foam cells been demonstrated in the histological specimens, and then in small quantities only. There is formation of new bone simultaneously with bone destruction in the osseous regions involved, and the lesion demonstrates segmental distribution in the skeletal system.

According to HOLT & WRIGHT (11) v. Recklinghausen's neurofibromatosis involves the skeletal system far more frequently than previously supposed, and several bones may be attacked. The roentgenological picture presented by the osseous manifestations of this lesion may resemble polyostotic fibrous dysplasia, though as a rule it has a more cystic appearance. In the great majority of cases of neurofibromatosis cutaneous fibromas may be demonstrated, and further, café-au-lait coloured pigment changes in the skin, which are in no way similar to the pigmentation of the skin in Albright's syndrome; elephantiasis frequently is found.

COLEY & STEWART (2) have observed two cases of polyostotic fibrous dysplasia with development of sarcomata (spindle and giant cell sarcomata). This development appears to be very rare.

In all the cases in this series the roentgenological picture has been so typical that the diagnosis has been easy. The diagnosis of polyostotic fibrous dysplasia can only be established on the findings of the roentgenological examination.

SUMMARY

The author gives a brief report of seven cases of polyostotic fibrous dysplasia of bone observed at the Oslo University Hospital, Department of Roentgenology during the past decade. The chief points regarding the roentgenological symptomatology of the lesion, differential diagnosis and histology are dealt with.

ZUSAMMENFASSUNG

Der Verfasser gibt einen kurzen Bericht über 7 Fälle mit Dysplasia fibrosa polyostotica, die während der letzten 10 Jahre an der Röntgenabteilung des Universitäts-Krankenhauses in Oslo beobachtet wurden. Die hauptsächlichen Kennzeichen in der röntgenologischen Symptomatologie dieser Erkrankung, die Differential Diagnose und die histologische Erkennung werden besprochen.

RÉSUMÉ

L'auteur fait une brève revue des cas de dysplasie polyostéique fibreuse de l'os observés à l'Hôpital Universitaire d'Oslo dans le service de radiologie au cours de la dernière décade. Les points essentiels de la symptomatologie radiologique de cette lésion, sa délimitation envers les autres maladies et son histologie sont décrits.

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FROM THE ROENTGEN DEPARTMENT (DIRECTOR: PROFESSOR OLLE OLSSON)
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A COMPLICATION OF PERCUTANEOUS CAROTID ANGIOGRAPHY[1]

by

Hans Idbohrn

In the Roentgen Department of the University Clinics of Lund carotid angiography is carried out exclusively by the method described by LINDGREN (1947).

Untoward reactions to this method of examination are fairly infrequent and may be due to the contrast medium itself or to the technique of the percutaneous puncture. The frequency and nature of complications observed in the Roentgen Department of the University Clinics of Lund were described by OLLE OLSSON in 1949. Hematomas following the puncture are common but usually not important complications. As might be expected, small hematomas are quite usual by reason of the fact that large-sized needles, permitting a more rapid injection of the contrast medium, are used. Immediately after the angiographic examination a film is taken in the ward, of the patient's neck and thorax in order to obtain some idea of the size and location of any hematomas present. If a hematoma be large enough to compress the trachea, or if it extends down into the mediastinum, it becomes of clinical importance. Hematomas, however, rarely assume such proportions. They cause as a rule only a slight tenderness of the neck and a very small rise in temperature.

If the end of the needle be ground very obliquely, whereby part of the tip lies outside the lumen of the vessel, or if the tip inadvertently slip through or out of the vessel during injection, the contrast medium will be deposited extravasally. This will cause pain and marked tenderness to palpation in the neck.

A complication of another kind due to puncture technique occurred in one case during percutaneous carotid angiography. This complication

[1] Submitted for publication, April 25, 1951.

was of a type giving such clinical symptoms and roentgen signs, that it is considered worthy of reporting, particularly as percutaneous carotid angiography is a method now being used with increasing frequency.

Case Report

A man, 40 years old (Neurosurg. 45/50), previously generally healthy, had been treated 6 months earlier for epilepsy. The patient was commencing to feel drowsy and to suffer from increasingly severe headaches, nausea and expressive aphasia. He was admitted to the Neurosurgical Clinic, Lund, with a suspected cerebral tumour.

Condition of nervous system on admission: slow cerebration of fairly marked type. Expressive aphasia. Paresis of the right facial nerve, of central type. Ophthalmological examination: slight bilateral papilledema (less than 1 diopter).

Roentgenograms of the skull showed no signs of any pathological condition.

Left carotid angiography was carried out on 19. 1. 1950. The left common carotid artery was punctured at its bifurcation. After the examiner had convinced himself that the tip of the needle was in the lumen of the vessel, 10 ml of the contrast medium was injected and the angiograms obtained. Before the injection the patient was somnolent but had not lost consciousness. A few minutes after injection his condition suddenly deteriorated, respiration became laboured and irregular and he lost consciousness. Oxygen was administered, and after about 10 minutes the patient recovered consciousness: his general conditions then became the same as before the injection.

On examination of the roentgenograms it was observed that most of the contrast medium had been deposited in the wall of the common carotid artery and caused a semilunar bulge inwards of the innermost coat just above the bifurcation: this narrowed the lumen considerably, particularly at the origin of the internal carotid artery (Fig. 1). The part of the arterial wall thus separated off was about 0.5 mm thick. Only a small portion of the contrast medium had been deposited into the lumen: it had disappeared from there but had been carried further away at an unusually slow rate. Even 4 seconds after the injection, by which time the contrast medium has usually passed into the veins, it was still to be seen in the finer branches of the arteries. The contrast medium was also still visible *in the vessel wall* around the site of the injection (Fig. 2), at and slightly above the bifurcation of the carotid artery.

On account of the reaction of the patient the examination was interrupted, but as it was considered important to make a diagnosis as soon as possible, it was repeated the following day.

Left carotid angiography on 20. 1. 1950. The common carotid artery was punctured fairly far down in the neck. At the level of the site of injection a filling defect in the otherwise filled vessel was now apparent. The upper margin of the defect was smooth, but below it was seen to be irregular and shaggy. Some of the contrast medium had been deposited into the middle of the filling defect, which was located in an area where the vessel wall was already markedly thickened (Fig. 3). The cerebral circulation was also severely retarded on this occasion. The anterior cerebral artery was displaced slightly to the right, and pathological vessels, contained in an area 4 cm in diameter, were visualized in the lower parts of the left parietal and occipital regions, deep in the hemisphere.

Roentgen diagnosis: malignant glioma + hematoma in the wall of the left common carotid artery following puncture.

At this examination no appreciable untoward reactions were observed. The patient's condition remained unchanged.

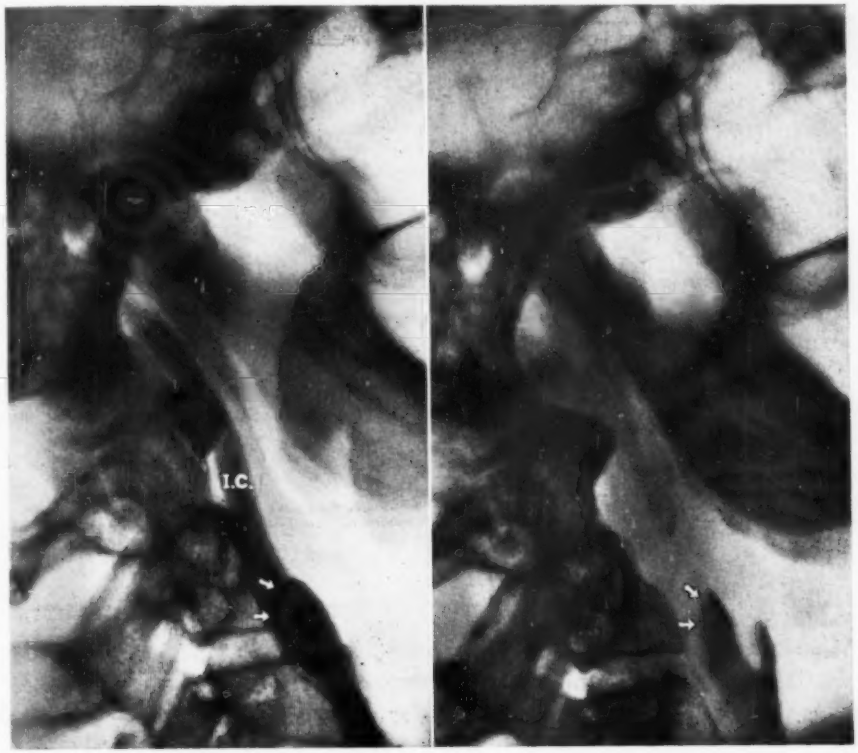

Fig. 1. Fig. 2.

Fig. 1. 19. 1. 50. Lateral view. A large portion of the contrast medium deposited into the wall of the common carotid artery forces the innermost layer of the vessel inwards and thereby considerably narrows the lumen of the internal carotid artery at the level of the bifurcation. Only a small amount of the contrast medium passed up into the internal carotid artery. The arrows indicate the site of intimal separation.

Fig. 2. Lateral view, 4 seconds after injection. Some of the contrast medium is still evident in the vessel wall, while the remainder has passed up into the internal carotid artery at an unusually slow rate.

On 24. 1. 1950 *diagnostic cerebral puncture* was performed (LUNDBERG).

Pathohistological diagnosis: malignant glioma.

The patient gradually became worse, and he died on 4. 2. 1950.

Postmortem examination (4. 2. 1950). There were signs of a fairly extensive atheromatosis with the presence in some areas of coalescent plaques in the arterial walls in the region of the bifurcation of the left common carotid artery. The changes were most advanced at the bifurcation and in the proximal part of the internal carotid artery. From

Fig. 3. Fig. 4.

Fig. 3. 20. 1. 51. Lateral view. At the site of the first injection a filling defect — hematoma — is now seen in the vessel wall.Circulation in the internal carotid artery is still retarded. A small amount of contrast medium was deposited into the middle of the hematoma. The arrows indicate the part of the margin of the hematoma facing the lumen.

Fig. 4. Preparation of the common carotid artery and its main branches. The arrow indicates the site of injection. The intramural hematoma is about 5 cm long. Fairly marked atheromatosis is evident.

the site of the injection, about 5 cm inferior to the bifurcation and extending to the origin of the internal carotid artery there was an *intramural* hematoma almost 6 cm long, 1 cm wide, and at the most, half a centimeter deep, located under the intima of the vessel wall. The lumen of the vessel was narrowed considerably at this level, particularly at the origin of the internal carotid artery.

On the right side the atheromatous changes in the carotid artery and its main branches were slightly less advanced.

Microscopic examination of sections from the common carotid artery showed a moderate degree of intimal atheromatosis. A relatively large, caudally tapering hematoma with thrombus formation was seen roughly in the middle of the media of the vessel on the left side. The outer layer of the media of that part of the carotid artery nearest the hematoma contained numerous polymorphonuclear leukocytes. In sections from the lower

part of the changed area (in the intimal defect) the defect was filled by a fresh white thrombus.

Pathological report: large multiform glioblastoma in the left hemisphere + mild to moderate general atheromatosis + hematoma with thrombus formation in the wall of the left common carotid artery.

Discussion

Carotid angiography is a very important examination method in suspected intracranial expansive formations. The percutaneous injection of a diagnostic contrast medium is now widely employed and reports of serious complications are therefore presumably justified and desirable.

In the case described practically the entire tip of the injection needle lay within the actual vessel wall. Most of the contrast medium was therefore deposited under pressure into the wall, so that a layer of the latter — mainly the intima — was separated and pressed inwards towards the lumen, which was thereby partly occluded in the same manner as by a thrombus at the origin of the internal carotid artery. A small portion of the contrast medium deposited into the lumen passed this level and was transported by the blood stream up into the anterior and middle cerebral arteries, but at a considerable reduced rate. It may be assumed the loss of consciousness was due to the retarded circulation, possibly in connection with impaired permeability of the cerebral vessels due to the increased time of administration of the contrast medium and to pathological changes in the walls of the vessels supplying the cerebral tumour (Broman & Olle Olsson 1948, 1949; Broman, Forssman & Olle Olsson 1950).

The intramural interstice produced in the vessel by the injection of the contrast medium became filled with blood and the resultant hematoma visualized in the angiograms carried out the following day gave a filling defect with a smooth superior and lateral outline but with a ragged inferior margin. At postmortem examination 16 days after the first injection a fairly long, caudally tapering hematoma with thrombus formation was seen in the media of the vessel wall. Moderate intimal atheromatosis possibly partly responsible for the separation of the vessel coats and for the wide spread of the hematoma was also noticed. It should be observed that the carotid sinus and bifurcation are the most frequent and earliest sites of arteriosclerosis of the nodular type, while diffuse arteriosclerosis is commonest in the arteries of the extremities (von Hasselbach 1931 and Keele 1933). The early and severe manifestation of arteriosclerosis of the carotid bifurcation and in the region of the sinus may possibly be explained by small traumata in the area of the carotid sinus and consequent injury to the intima (Sponer 1942). The continuous fluctuation in the blood pressure in the sinus due to its function as a

regulator of the blood pressure may overburden its walls with resultant increased tendency to vascular changes (SORGO 1939).

A few cases are on record in which thrombosis was believed to originate at the site of the injection into the artery. NORDMANN (1937) reported a case in which an embolus of the middle cerebral artery following carotid angiography was thought to have formed at the site of the puncture in the carotid. EKSTRÖM & LINDGREN (1938) described a similar case. HULTQUIST (1942) observed a small thrombus at the site of the puncture in a case following angiography. He remarks, however, that a fragment of the tip of the injection needle might have been a stimulating or contributory cause of the thrombosis. No earlier reports of a complication such as that described above are found in the literature.

SUMMARY

Some of the contrast medium injected percutaneously for the purpose of carotid angiography was deposited under pressure into the wall of the artery. The inner layer of the vessel was thereby separated with a resultant hematoma which bulged into and narrowed the lumen of the artery causing consequent loss of consciousness. Postmortem examination after death from malignant glioma confirmed the roentgen diagnosis.

ZUSAMMENFASSUNG

Bei perkutaner Angiographie der Carotis kam es zu einem Druck-Infiltrat von etwas Kontrast-Mittel in der Wand der Arterie. Dabei kam es zu einer Ablösung der inneren Wandschicht des Gefässes und zur Entstehung eines Hämatoms, das sich nach innen vorwölbte und das Lumen der Arterie einengte mit anschliessendem Verlust des Bewusstseins. Nach erfolgtem Tod an malignem Gliom bestätigte die Obduktion die Röntgen-Diagnose.

RÉSUMÉ

Une partie de la matière contrastante injectée à travers la peau au cours d'une angiographie de la carotide a été déposée sous pression dans la paroi même du vaisseau. La couche intérieure fut ainsi détachée et il en résulta un hématome qui faisait saillie et qui comprimait la lumière artérielle entraînant la perte de la connaissance pour le malade. Un gliome malin ayant provoqué la mort, l'autopsie confirma le diagnostique radiologique.

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ON ROENTGENOGRAPHY AT 200 kV[1]

by

K. Lindblom

The large skin doses administered in roentgen kinematography and the long exposure times required for the gastro-intestinal roentgenography of thick patients were the two main reasons for experiments with techniques at 200 kV peak.

JANKER, already in the thirties, constructed an apparatus for roentgen kinematography at 125 kV.

For the further development of roentgen kinematography it is important to diminish the amount of radiation, at least on the skin, to a minimum. Experiments for this special purpose were carried out in 1945. A roentgen therapy apparatus was used for roentgen examinations of the stomach and the colon. The results of these experiments stimulated the Järnhs Laboratories in Stockholm to build a four-valve apparatus for exposures at a maximal voltage of 200 kV. In May 1950 this apparatus was installed at the Karolinska Sjukhuset. The mA capacity is 100 at 200 kV but as the roentgen tube at present in use has no rotating anode the total effect live load cannot be used. The Machletts Laboratories have however held out the prospect of building an adequate rotating anode tube.

The results with the apparatus have demonstrated that it is possible to use 200 kV for roentgenography without too much loss of contrast in the roentgenograms but with a gain of a considerable decrease in the amount of radiation.

Figs. 1—2 illustrate the roentgenograms of a stomach examined at 200 and 100 kV respectively. Focus-film distance, screens, film and development were the same in the two films, a single Lysholm grid was used in both exposures. The 100 kV film is taken at 20 mA sec. and the 200 kV film at 3 mA sec.

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Fig. 1.

Fig. 2.

In roentgen kinematography raising the voltage from 100 to 200 kV means decreased radiation of the tube-side of the skin to less than 1/8, provided the same focus-film distance is used. In routine gastro-intestinal work, for example, the 200 kV will enable the exposure time enough to be shortened for movements to be avoided. As the decrease in exposure time is more than is necessary for this purpose, there is a hypothetic possibility of also increasing the sharpness, by diminishing the focus spot of the tube.

The contrast in the roentgen pictures at 200 kV is distinctly less than in pictures at a lower voltage. However, if the 200 kV apparatus be combined with the MORGAN television set the lack of contrast can be easily compensated in the television apparatus.

SUMMARY

With an apparatus for 200 kV it is suggested that the rise in voltage is useful and valuable in diagnostic roentgenology.

ZUSAMMENFASSUNG

Mit einem Apparat für 200 kV wird demonstriert, dass eine solche Steigerung der Spannung für diagnostische Röntgenologie verwendbar und wertvoll ist.

RÉSUMÉ

L'auteur utilise pour le radiodiagnostic un appareil de 200 kV. Il montre que de tels voltages sont utilisables et ne sont pas dénués d'intérêt en radiodiagnostic.

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HEPATIC VENOGRAPHY[1]

by

A. M. Rappaport

With the technical assistance of WALTER COWAN

The radiography of the liver is still an unsolved problem. The various methods devised to make the liver parenchyma visible by saturating its cells with a radiopaque substance have not come into use (OLSSON and EKMAN). On the other hand the absorption of a radiopaque substance might be expected to be defective in the cells of a diseased organ. Also their use has been shown to be not without danger even for the normal liver cell.

In 1929 colloidal thoriumdioxyd was introduced by OKA, RADT, as a radiopaque substance in radiology. For many years the injection of this substance was practised to increase the density of the shadow cast by the liver parenchyma by making the v. Kupffer cells absorb the radiopaque material. However, the blockage of the reticulo-endothelial apparatus in liver and spleen was observed to be permanent and has proven to be noxious (HANS POPPER, NAEGLI, LANARI, JACOBSEN, and others). The enthusiasm about the hepatolienography with Thorotrast (25 % thoriumdioxyd) has faded away during the years 1929—1939 and the procedure is now in complete disuse. Other methods for the radiology of the liver deal with the visualization of the greater bile ducts and give no further information about the parenchyma itself.

The arteriography of the liver as the result of thoracic aortography is obtained as an inconstant filling of the hepatic artery with radiopaque substance, in cases where the site of the puncture of the aorta is close to the celiac axis. However, WAGNER et al. reserve thoracic aortography for cases in which »simpler and standard studies have failed to yield sufficient information».

Abdominal venography was first described by DOS SANTOS and applied by FARINAS, B. J. O'LOUGHLIN, and others. It is a further elabo-

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ration of the venography introduced by SGALITZER, WOHLLEBEN, and others. This abdominal venography renders only the great abdominal veins visible. It marks, however, the site where the hepatic veins branch off without giving further information about these veins.

The study of the vascular tree of the hepatic veins lies in the favourite field of the anatomist. H. N. SEGALL, using the technique of GROSS, obtained radiographs of the hepatic veins from liver specimen. The plates reveal a dense tuft-like arborization of the liver veins throughout the whole organ. Venous branches of seventh order can be observed. They join each other at angles of about 80—100°. The larger radicals of second to third order converge more acutely, forming angles of 20—45° at their junctions. It is suggested that anomalies in these regularly arborizing veins might reveal pathological changes in the liver substance.

To inject the hepatic veins in vivo with a radiopaque substance, a catheterization of these veins is necessary. The latter has become a routine procedure in humans since BRADLEY, INGELFINGER and associates have introduced their method of estimation of hepatic blood flow. Usually, they were able to catheterize only the right hepatic veins and they had to use cardiac catheterization. The method presented here deals with a guided catheterization of all liver veins in the dog.

Method

A transflex tubing, French No. 10, arranged after the principle of a Foley catheter, and having at its end a metal cap, is guided by an inside metallic leader (Fig. 1 A), the distal end of which can be curved in one direction at the command of a nut (n). The direction in which the curved leader points is indicated by the little screw (s). The transflex catheter, armed with the straight leader (A) is introduced through the saphenous vein (in smaller dogs through the femoral vein) into the vena cava until it passes the diaphragm. Under the fluoroscope the leader is curved by turning the nut. The curve is directed towards the right or left, depending on the intention to catheterize the right or left liver veins. By pulling back catheter and curved leader as a whole, the smooth metal cap of the curved catheter slides along the slightly stretched wall of the inferior vena cava until it falls into the mouth of a hepatic vein (Fig. 1 B). At this moment the transflex catheter is slid over the leader and slips easily into the lumen of the hepatic vein (Fig. 1 B). Then the shadow of the metal cap wanders laterally and becomes separated from the tip of the curved leader. The leader is gently removed while its curve is gradually released. The metal cap of the catheter lies now horizontally in the liver shadow (Fig. 1 B and Fig. 2) lateral from the midline and shares the respiratory movements of the diaphragm

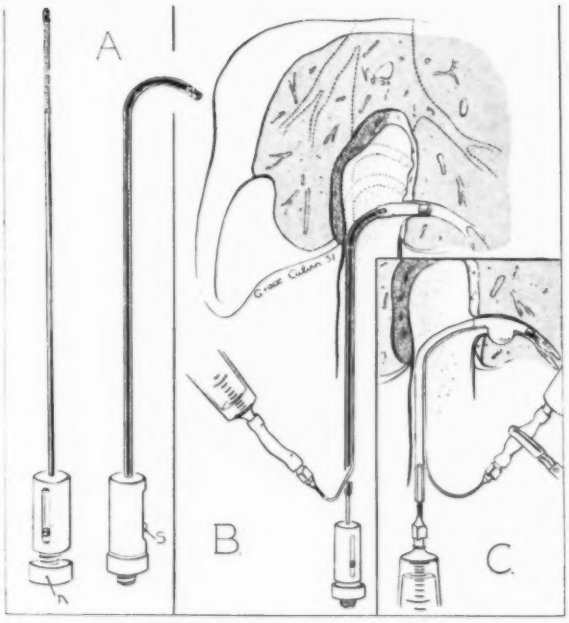

Fig. 1. Metallic leader and catheter.

A. In stretched and in curved position.
(n) nut commanding the curving of the leader.
(s) screw indicating the direction into which the curve of the leader points.

B. The curved catheter has entered a hepatic vein. The metal cap is separated from the upper end of the leader.

C. The catheter in situ. Its balloon, inflated and wedged into the hepatic vein, occludes this vessel temporarily.

and liver. A few cc of 35 % Diodrast are injected through the small tubing into the rubber balloon until it is well inflated (Fig. 1 C) and the catheter has become wedged into the catheterized vein (Fig. 2). Twenty cc of 70 % Diodrast are then injected through the catheter into the liver vein and three to four pictures, at one-half second intervals, are taken.

Following injection the corresponding part of the liver lobe is flooded with radiopaque substance. There is a short stagnation of flow in the liver lobe drained by the vein used for injection. Figure 3 shows the stage where a wedge-shaped part of the liver is intensely shadowed by the stagnant radiopaque substance.

Fig. 2. The balloon is inflated by injected radiopaque material which makes it visible in the liver shadow. Note the horizontal position of the metal cap at the end of the catheter.

Fig. 3. A wedge-shaped part of the liver is flooded with 70 % Diodrast.

Figure 4 shows the spread of the Diodrast into the rest of the liver substance through anastomotic channels with the other liver veins. The entire liver casts a darker shadow in which the liver veins become visible. It looks as if not only the nearest vascular channels but also the remotest ones are used to carry away the injected radiopaque substance. The Diodrast circulates through the whole liver and flows into the inferior vena cava through different hepatic veins.

Although the shadow-producing substance was injected into the right hepatic vein of a normal dog's liver, the vascular tree of the left hepatic veins becomes visible too and displays ramifications even of the fifth order. Thus the injection of Diodrast into one hepatic vein renders the vascular pattern of all liver lobes visible.

Figure 5 shows a dog's liver to which the blood supply was greatly reduced by previous operations. The stagnant radiopaque substance fills a right liver lobe which is smaller than normal.

Figure 6 shows (in spite of the kinked catheter) a good filling of the injected vein. Instead of the finer root-like arborizations of the vascular tree we observe here a sacculation of the venous branches. The liver veins in these atrophic livers are apparently deprived of their finer

Fig. 4. The vascular pattern of the right and left hepatic veins has become visible.

Fig. 5. Stagnant radiopaque substance fills an atrophic right liver lobe.

root-like affluents which help interconnect different vascular areas. The quick injection of Diodrast into these veins might have produced a temporary forceful dilatation of the branches of third order, due to a reduction of anastomosis in the atrophic liver lobe. Nevertheless the radiopaque substance has spread into the rest of the liver and produced a net delineation of the organ.

Figure 7, taken a second later, visualizes main branches of the hepatic venous tree which, however, lack regularity and distinctness as seen in Figure 4, the venogram of a normal dog's liver.

Conclusions

A method of visualization of the hepatic veins by direct injection of a radiopaque substance through a special catheter into a liver vein has been devised.

A new catheterization technique, using a metal leader which allows a guided catheterization of right and left liver veins, has been elaborated.

The visualization of the venous vascular tree of the liver in its finer ramifications gives a net delimitation of this organ from adjacent radiological shadows and reveals some pathological changes in the parenchyma of an atrophic liver.

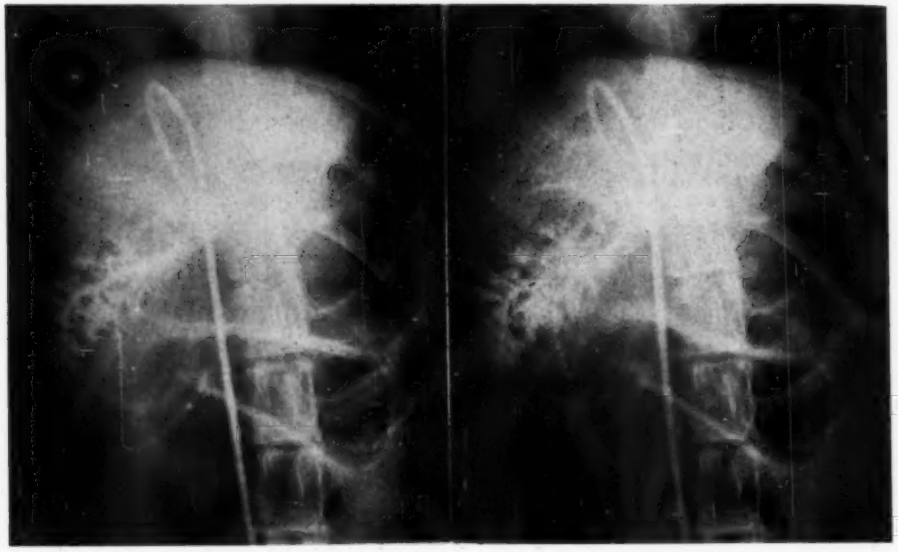

Fig. 6. Sacculation of the hepatic vein branches of the third order in an atrophic liver.

Fig. 7. The main branches of the hepatic venous tree are less visible than in a normal liver. Sharp delineation of the left liver lobe.

It is reasonable to suppose that specific changes produced in the vascular pattern of the liver, as, for instance, by tumours, abscesses, cysts, cirrhosis, etc., might also be disclosed, but these clinical studies have not as yet been attempted.

The author is very much indebted to Professor C. H. BEST for encouragement, and for the provision of facilities for this work.

SUMMARY

A new method of visualization of the hepatic veins by direct injection of a radiopaque substance through a special catheter into a liver vein is described.

ZUSAMMENFASSUNG

Es wird eine neue Methode zur Sichtbarmachung der Lebervenen beschrieben, bei der ein Kontrastmittel durch einen Spezialkatheter direkt in eine Lebervene injiziert wird.

RÉSUMÉ

Description d'une nouvelle méthode pour rendre visibles les veines hépatiques par l'injection directe d'une substance radio-opaque dans l'une des veines du foie avec l'aide d'une sonde spéciale.

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PRELIMINARY NOTICE

THE THIRD
SYMPOSIUM NEURORADIOLOGICUM

will be held in Stockholm on 17th—20th September 1952.

President: Professor ERIK LINDGREN, Stockholm

Vice-president: Professor OLLE OLSSON, Lund

Secretary: Docent ÅKE LINDBOM, Stockholm

Main Themes: Vertebral angiography
Roentgen diagnosis of spinal tumours
Phlebography in neuroradiology

Papers will also be read on various neuroradiological subjects.

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Radiologists, neurosurgeons and neurologists will be welcome at the Symposium. All those wishing to attend should send in their names and addresses to the Secretary. A detailed programme will be published early in 1952 in the same journals as this preliminary notice and also sent to those who have expressed their intention of being present. Arrangements for hotel accommodation in Stockholm will be announced in due course. The social programme will include plans for the entertainment of the ladies accompanying Members. The subscription fee will be approximately Sw. Kr. 75: — for active member and Sw. Kr. 25: — for companion.

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